Pediatric diffuse interstitial pneumonia: demonstration of 3 cases

Children's interstitial lung disease (chILD) encompasses a heterogeneous group of innate, genetic, infectious and inflammatory diseases, quite different from that seen in adulthood. ChILD1 has two clinical types: a) a perinatal progressive type, and b) a late-onset slowly progressive type. At least three of the following four criteria should be requested for the diagnosis of chILD. 1) respiratory symptoms (cough, rapid and/or difficulty breathing, exercise intolerance), 2) respiratory signs (resting tachypnea, adventitious sounds, retractions, digital clubbing, failure to thrive, respiratory failure), 3) hypoxemia, and 4) diffuse abnormalities on chest radiography or high-resolution computed tomography (HRCT). The following commoner disorders should be excluded, such as cystic fibrosis, acquired or congenital immunodeficiency, congenital heart disease, bronchopulmonary dysplasia, pulmonary infection, primary ciliary dyskinesia and recurrent aspiration. VATS sampling is recommended to evaluate microscopic features of child.

Three VATS-sampled cases of chILD (23-day-old, 3-month-old and 1-year old) are presented.

Ref.-1: Devine MS, Garcia CK. Genetic interstitial lung disease. Clin Chest Med 2012; 33(1): 95-110. doi: 10.1016/j.ccm.2011.11.001

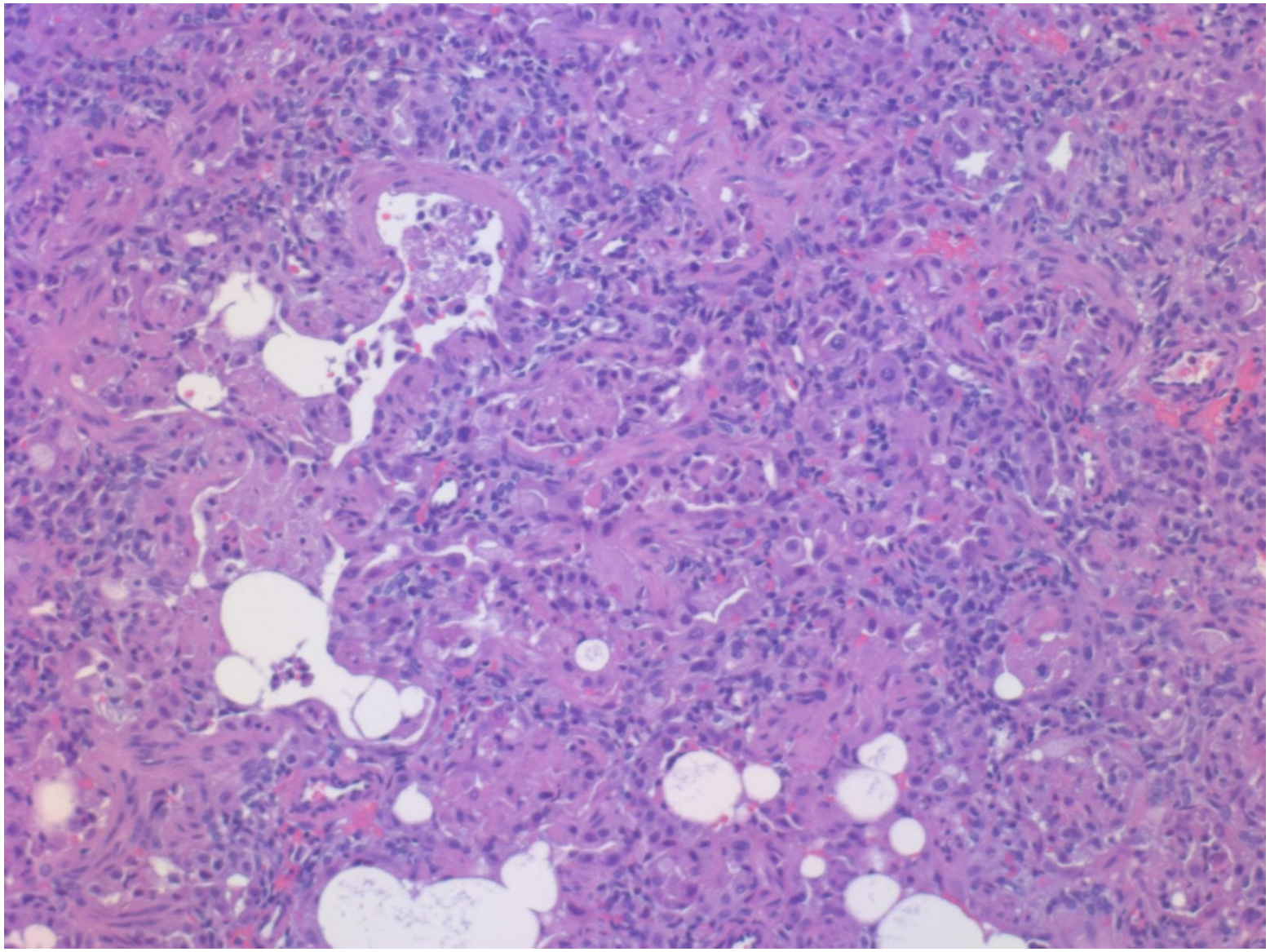
Ref.-2: Lee EY. Pediatric interstitial (diffuse) lung disease. Imaging Pediatr Pulmonol 2019; 25: 145-197. doi: 10.1007/978-3-030-23979-4_8

Ref.-3: Deutsch GH, et al. Diffuse lung disease in young children: application of a novel classification scheme. Am J Respir Crit Care Med 2007; 176(11): 1120-1128. doi: 10.1164/rccm.200703-393OC

Case 1 (a 23 day-old female newborn)

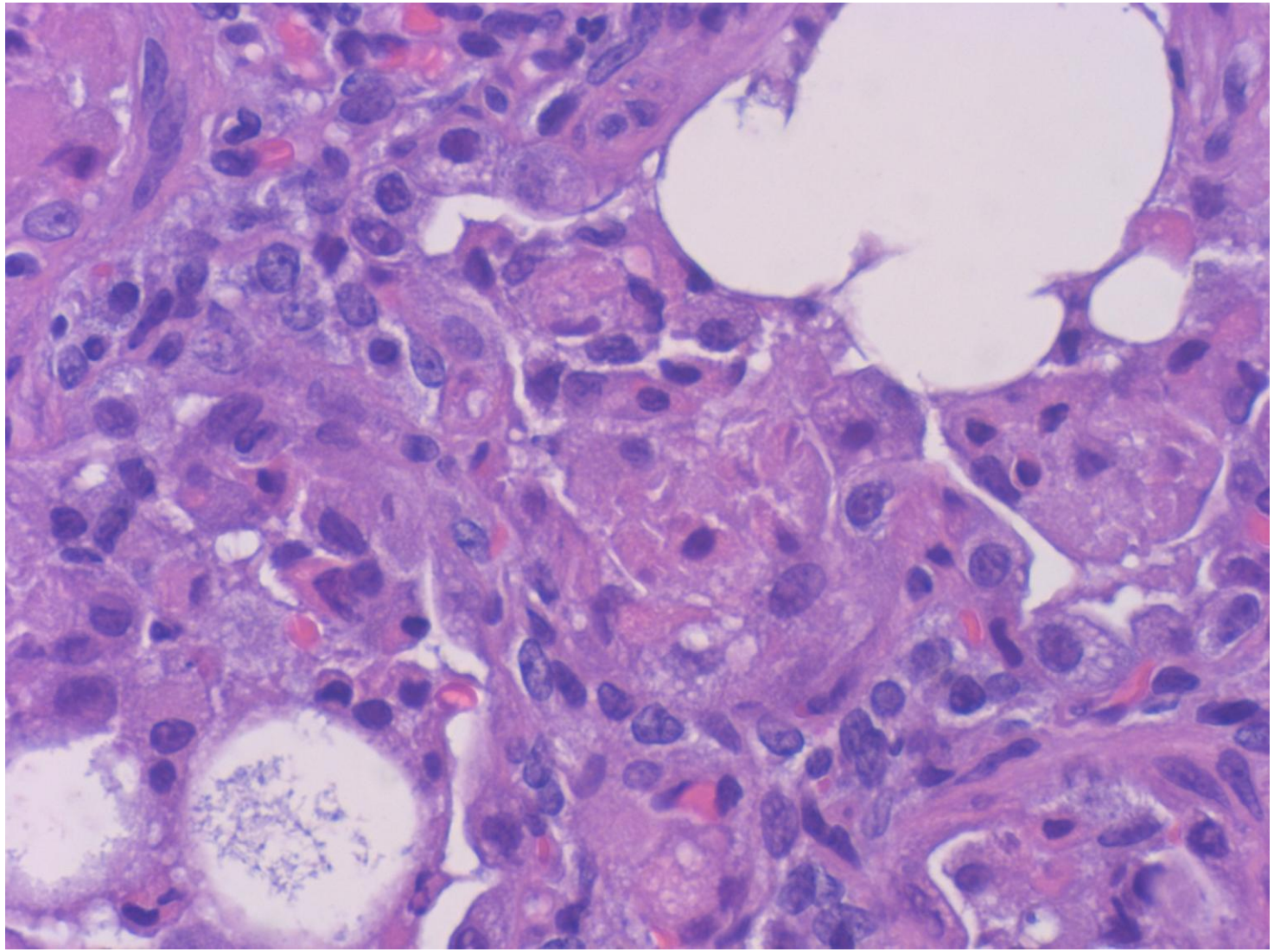
A girl was born at the 40th gestational week with body weight of 2,234 g. She immediately complained of respiratory failure with bilateral pneumothorax. After intubation, CPAP and antibiotics administration started. Chest imaging study reveals diffuse ground-glass opacity with KL-6 (Krebs von den Lungen-6): 968 U/mL. PSL was added, but poor respiratory function persisted. VATS biopsy was taken at the 23th day after birth.

Case 1



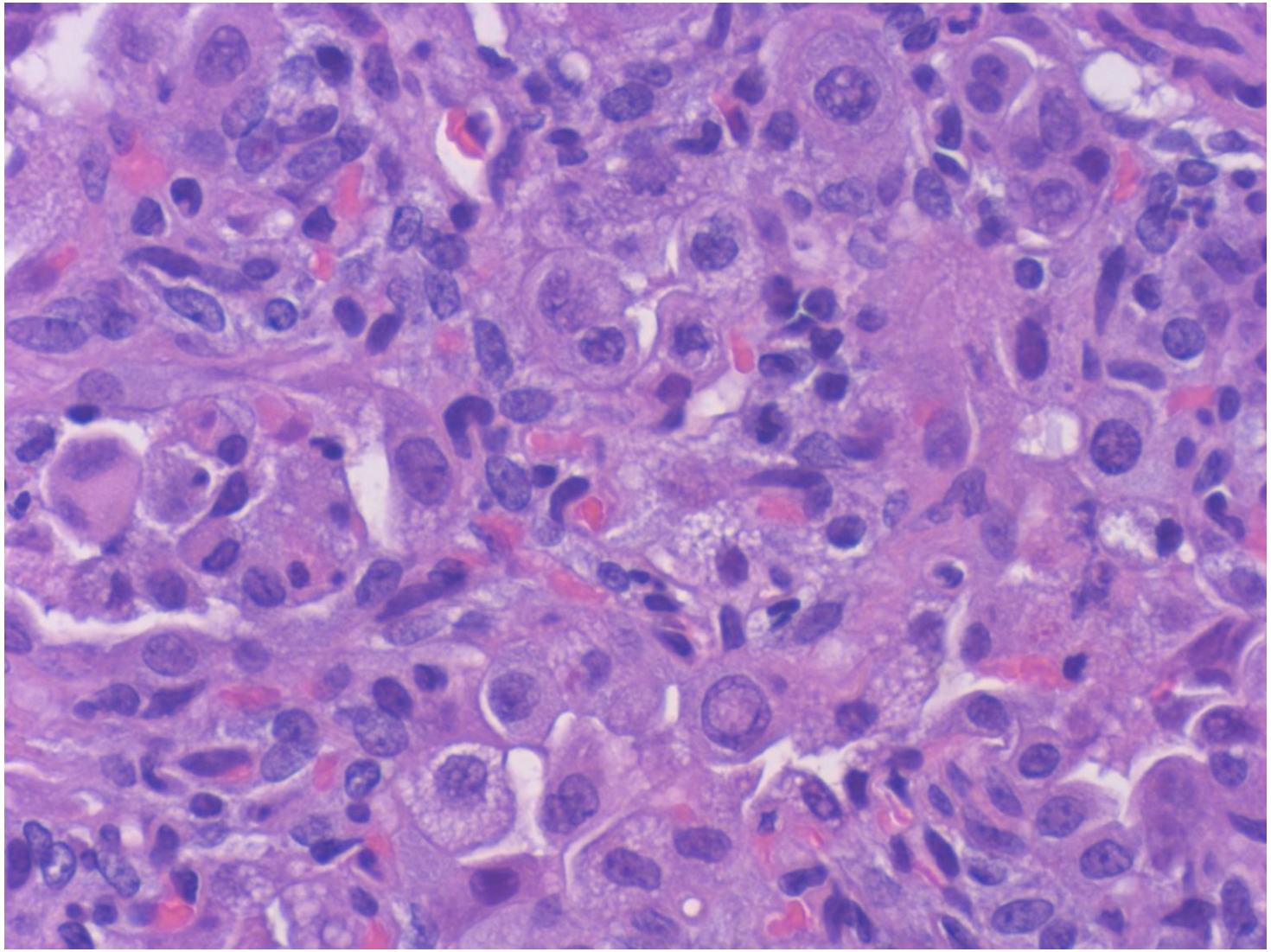
Hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, diffuse collapse of the alveolar lumina with activation of type II pneumocytes is evident. Accumulation of alveolar macrophages is multifocally associated. H&E-1

Case 1



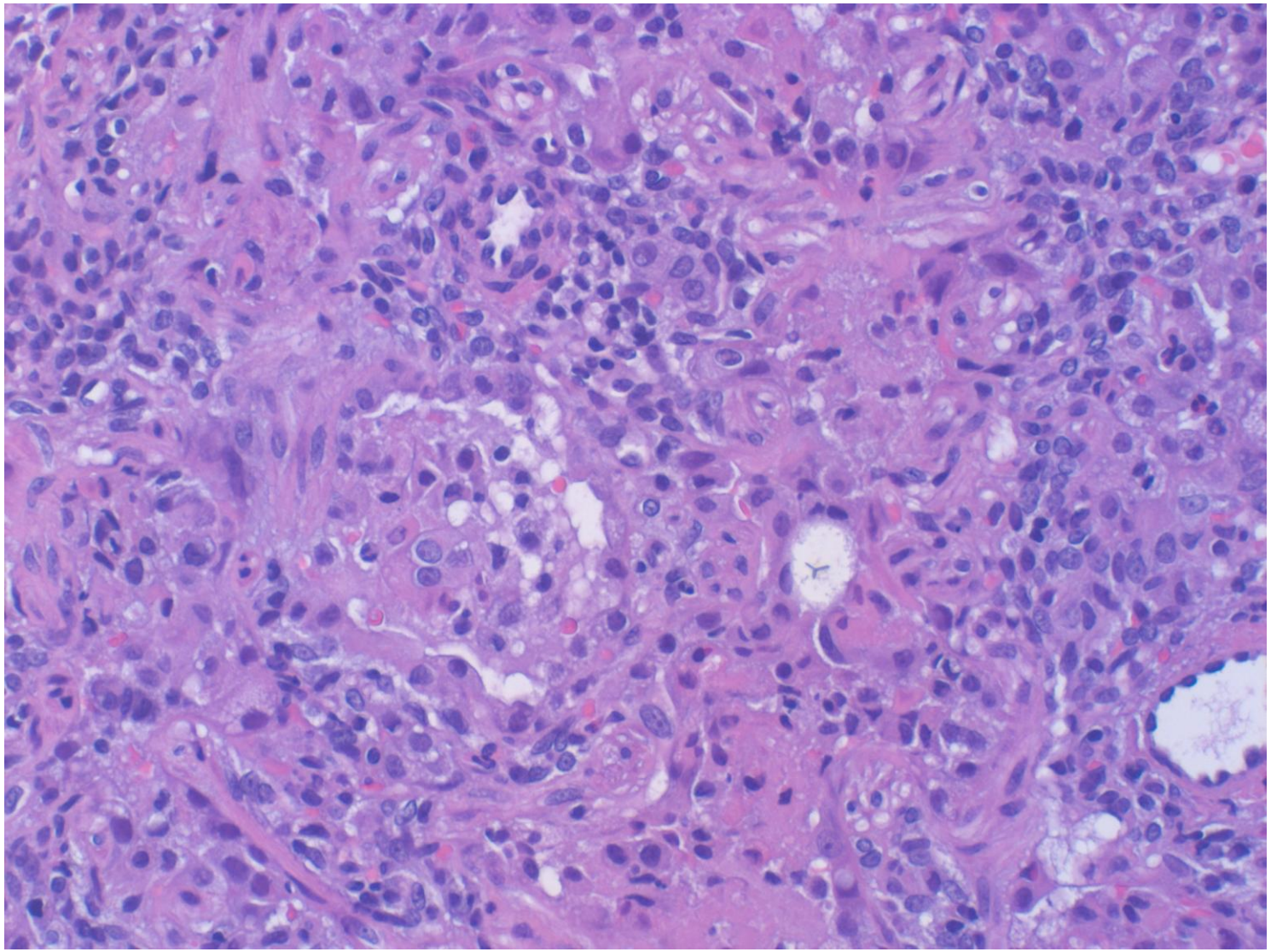
Hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, diffuse collapse of the alveolar lumina with activation of type II pneumocytes is evident. H&E-2

Case 1



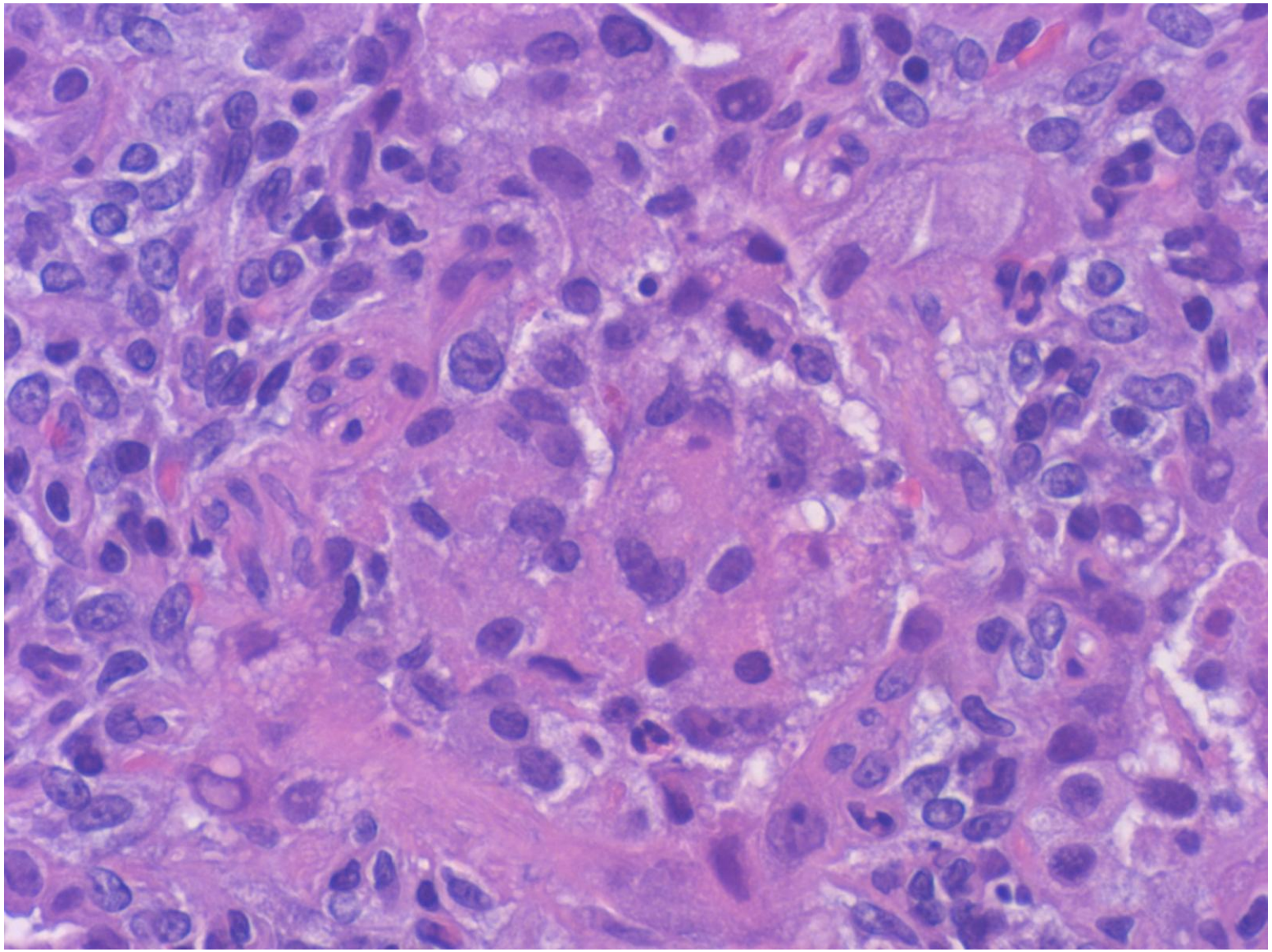
Hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, diffuse collapse of the alveolar lumina with activation of type II pneumocytes is evident. H&E-3

Case 1



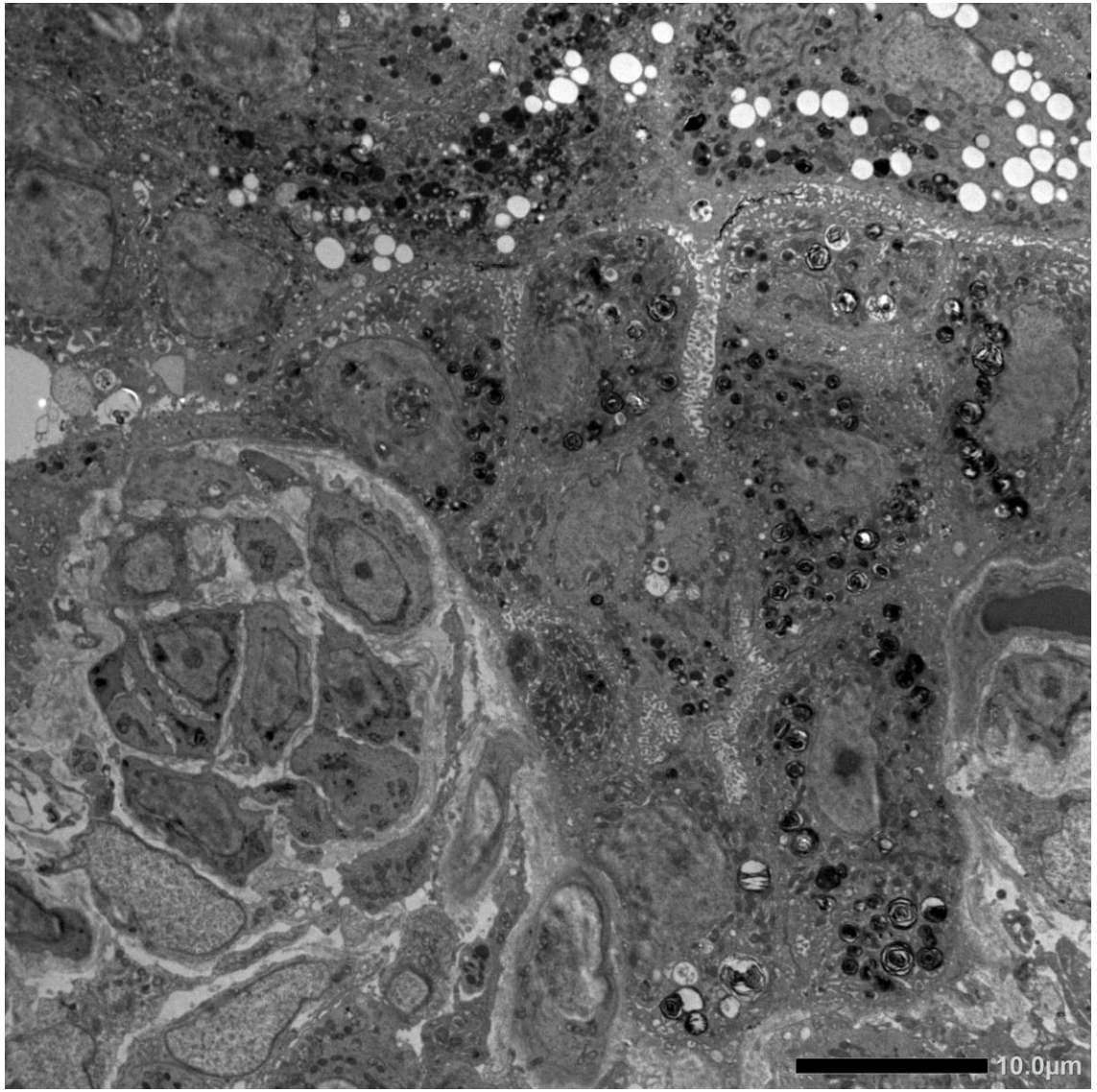
Hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, diffuse collapse of the alveolar lumina with activation of type II pneumocytes is evident. H&E-4

Case 1



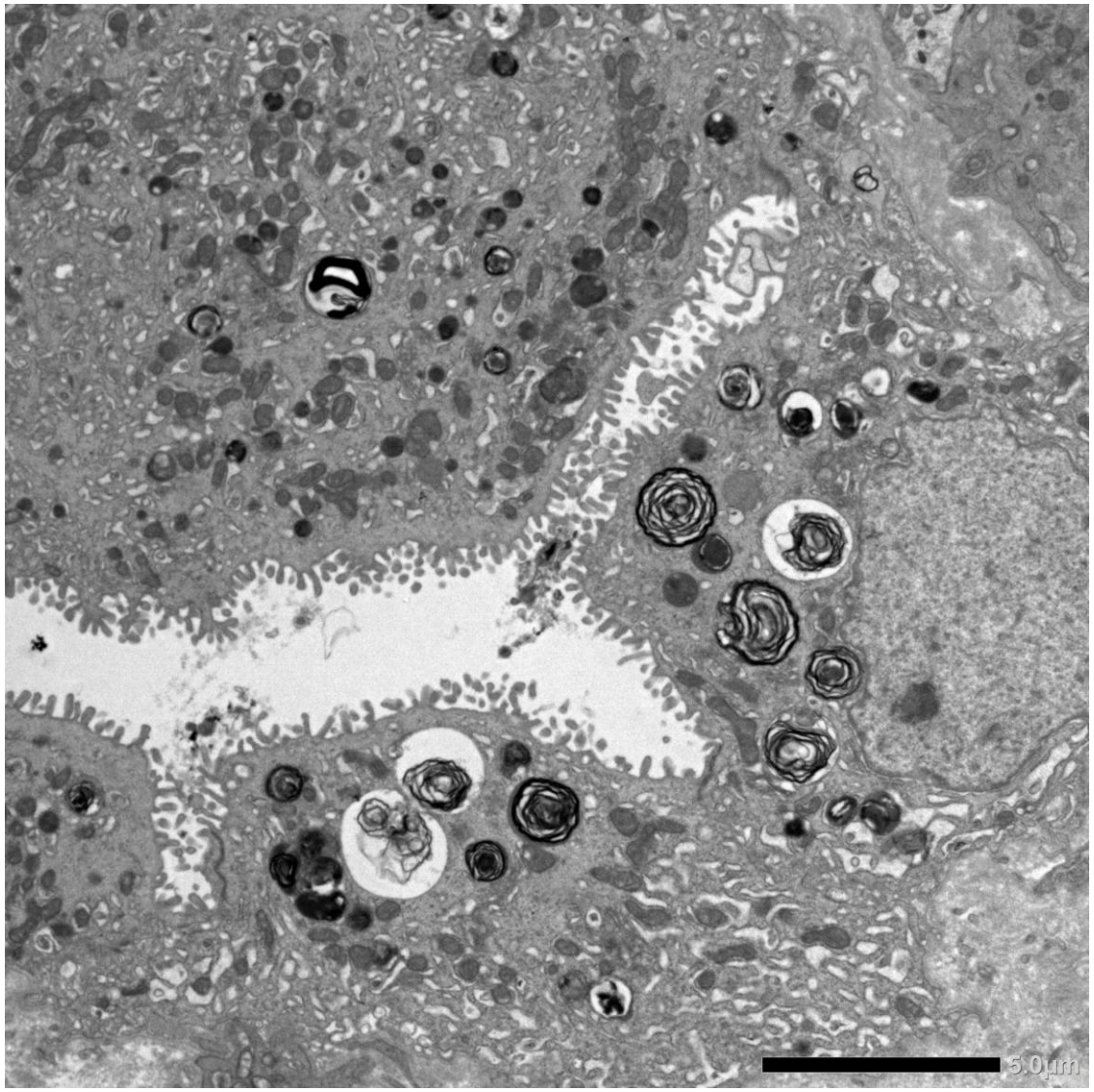
Hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, diffuse collapse of the alveolar lumina with activation of type II pneumocytes is evident. H&E-5

Case 1



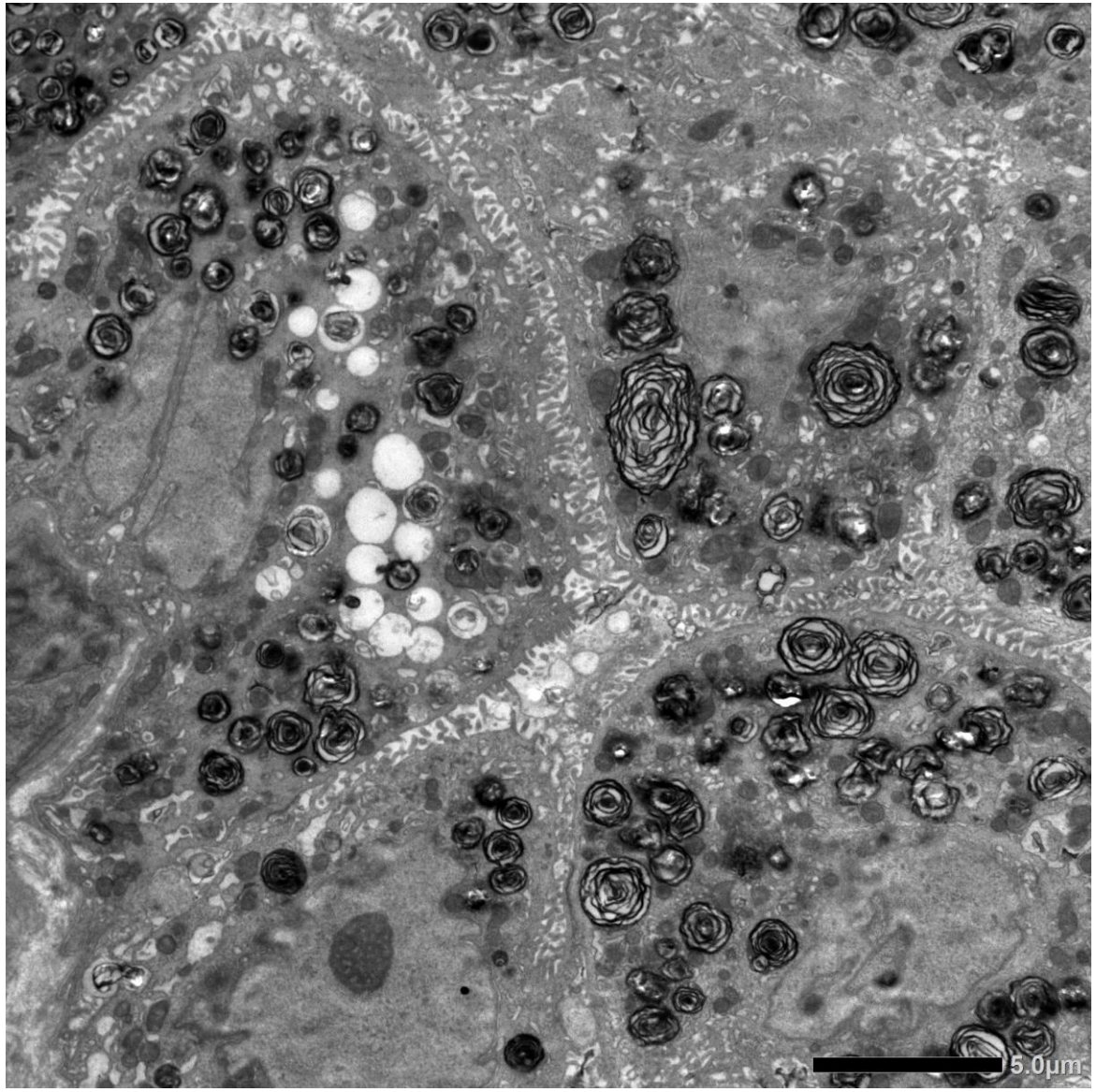
Ultrastructure of hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, activated type II pneumocytes with numerous lamellar bodies occupy the collapsed alveolar lumen. Accumulation of alveolar macrophages is also noted. EM-1

Case 1



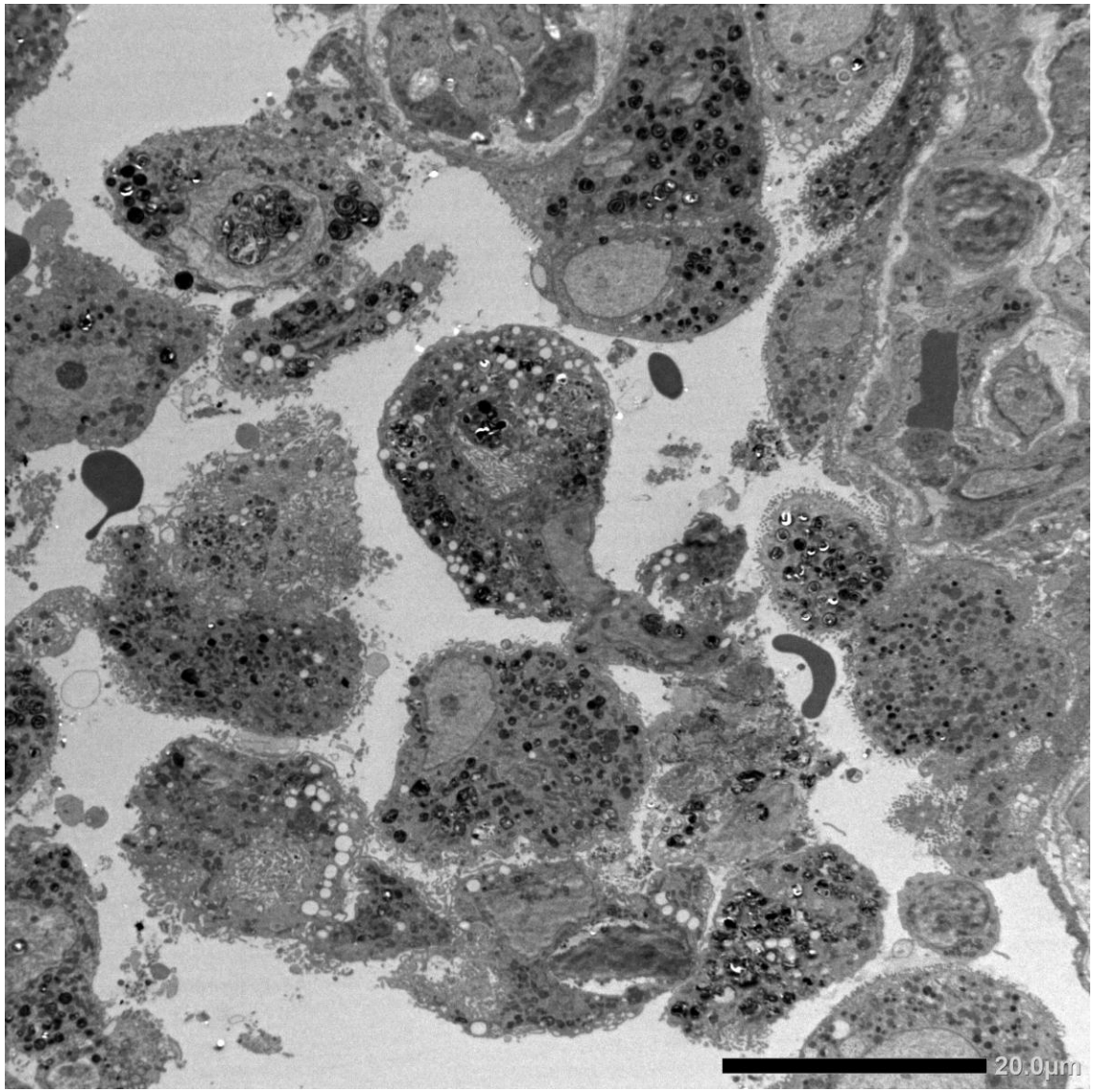
Ultrastructure of hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, activated type II pneumocytes with numerous lamellar bodies occupy the collapsed alveolar lumen. Microvillous surface is observed. EM-2

Case 1



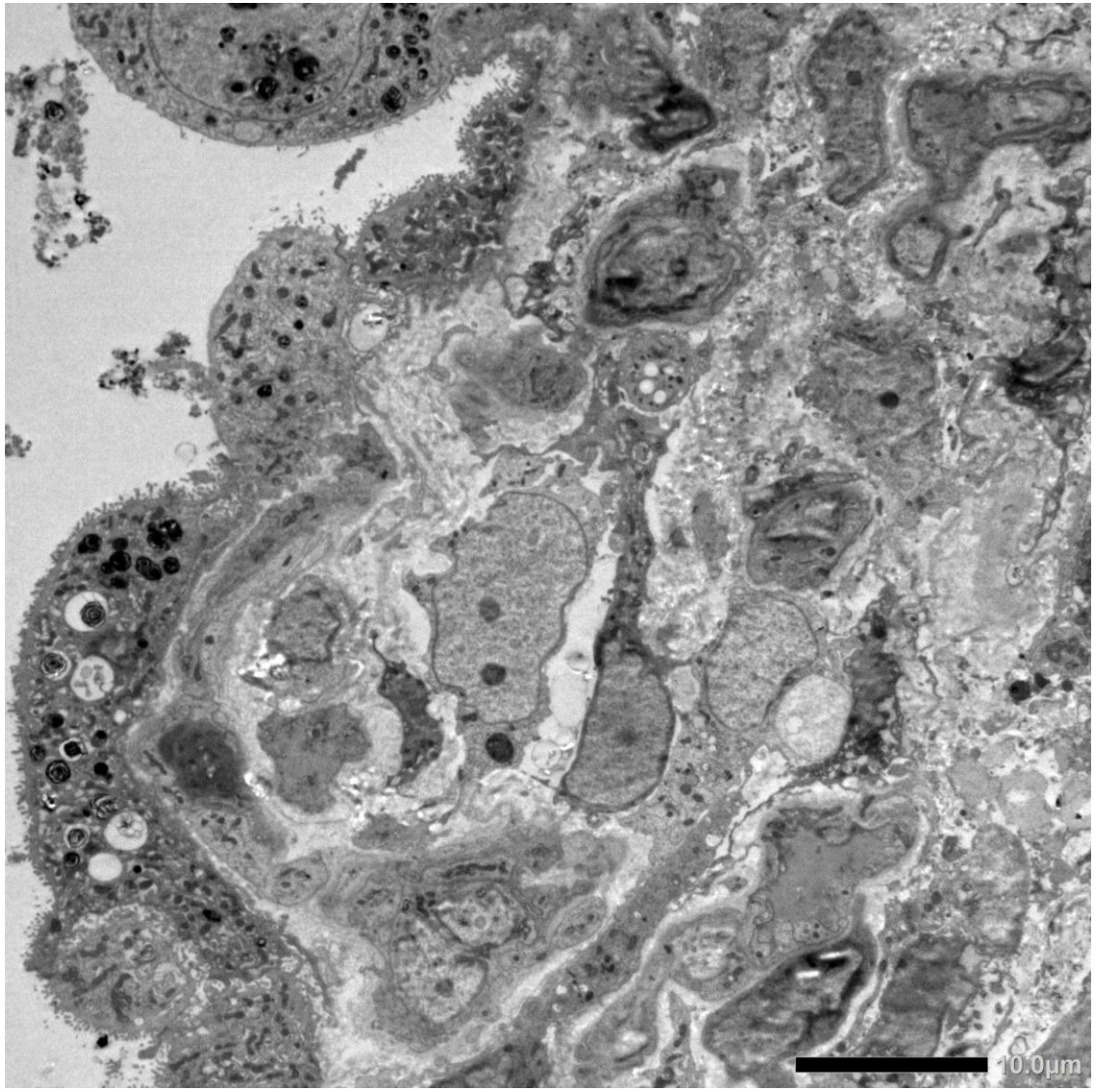
Ultrastructure of hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, activated type II pneumocytes with numerous lamellar bodies occupy the collapsed alveolar lumen. Microvillous surface is observed. EM-3

Case 1



Ultrastructure of hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, activated type II pneumocytes with numerous lamellar bodies are exfoliated into the alveolar lumen. EM-4

Case 1

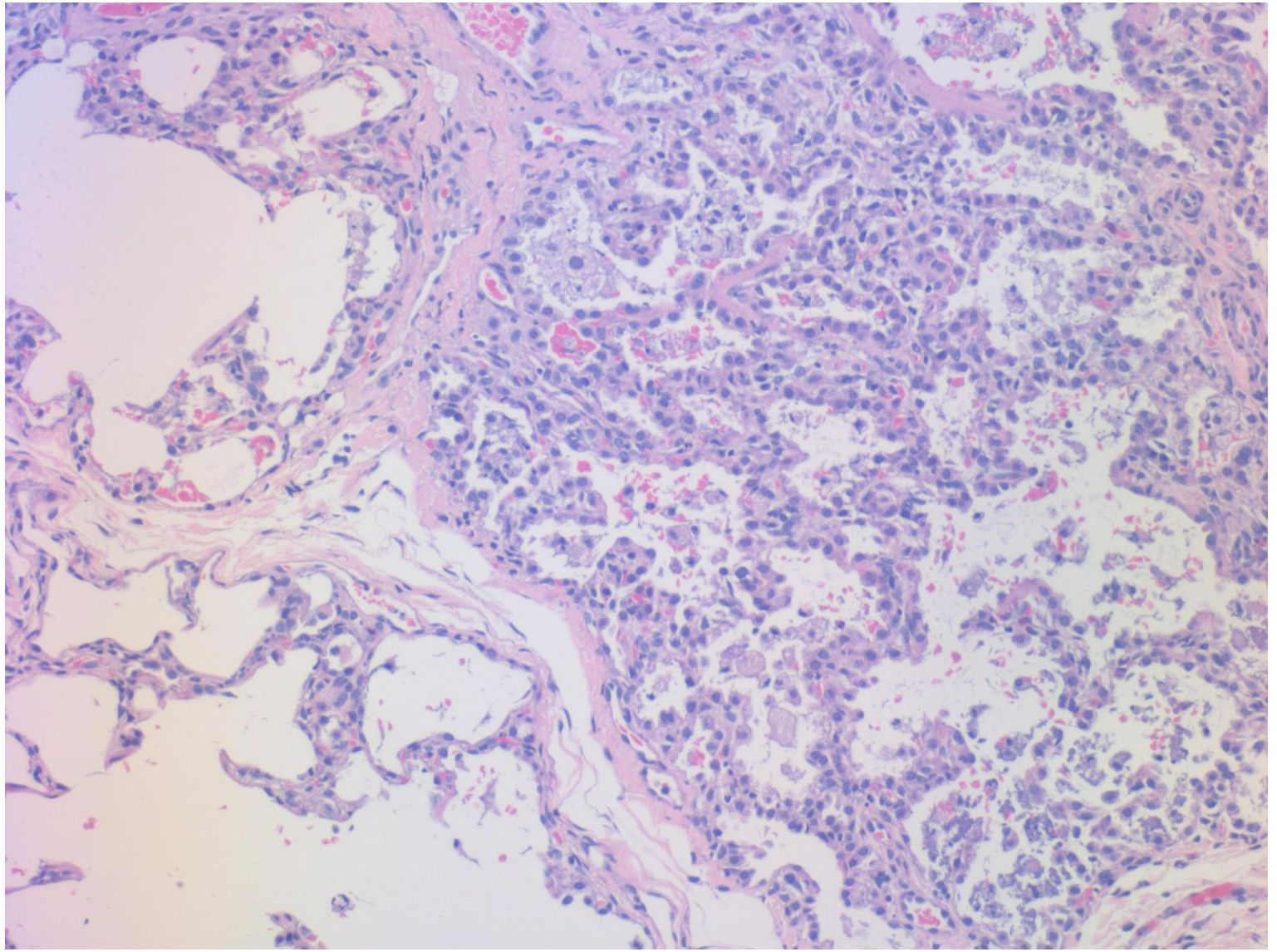


Ultrastructure of hereditary interstitial pneumonia seen in a 23-day-old female newborn. In this severe form of chILD, activated type II pneumocytes with numerous lamellar bodies are hyperplastic, and fibroblastic growth is associated in the interstitial tissue. EM-5

Case 2: A 3 month old girl with 21 trisomy

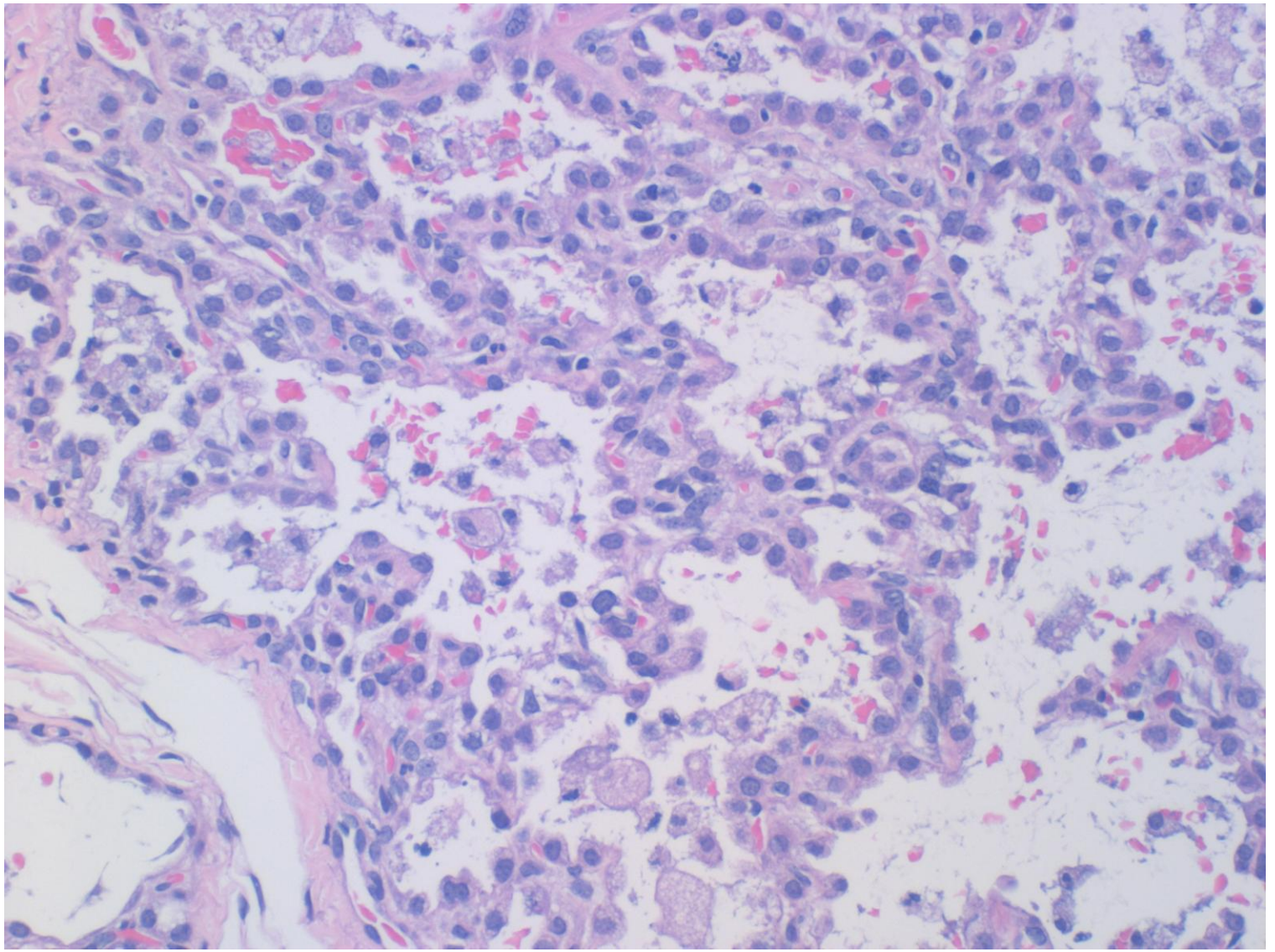
A female newborn was diagnosed as 21-trisomy (Down's syndrome) with patent ductus arteriosus and anal atresia, and surgical treatments were performed for both lesions. In the routine checkup at 3 months after birth, respiratory failure was pointed out. Chest CT suggested interstitial lung disease, and serum KL-6 was elevated. VATS biopsy was taken. chILD may be associated with chromosomal abnormalities.

Case 2



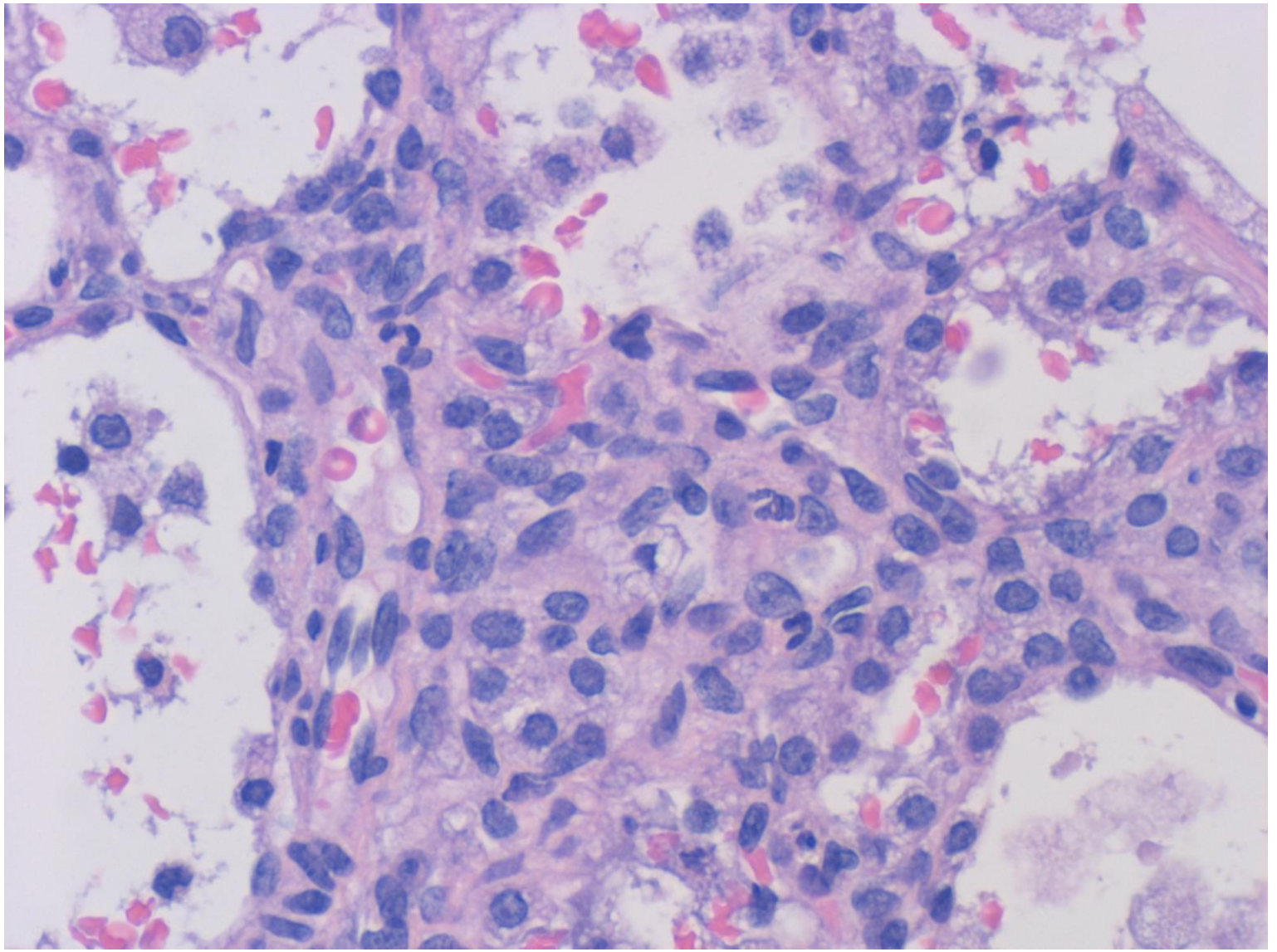
Interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activation of type II pneumocytes is seen along the mildly thickened alveolar septa. Exudation of alveolar macrophages is associated. H&E-1

Case 2



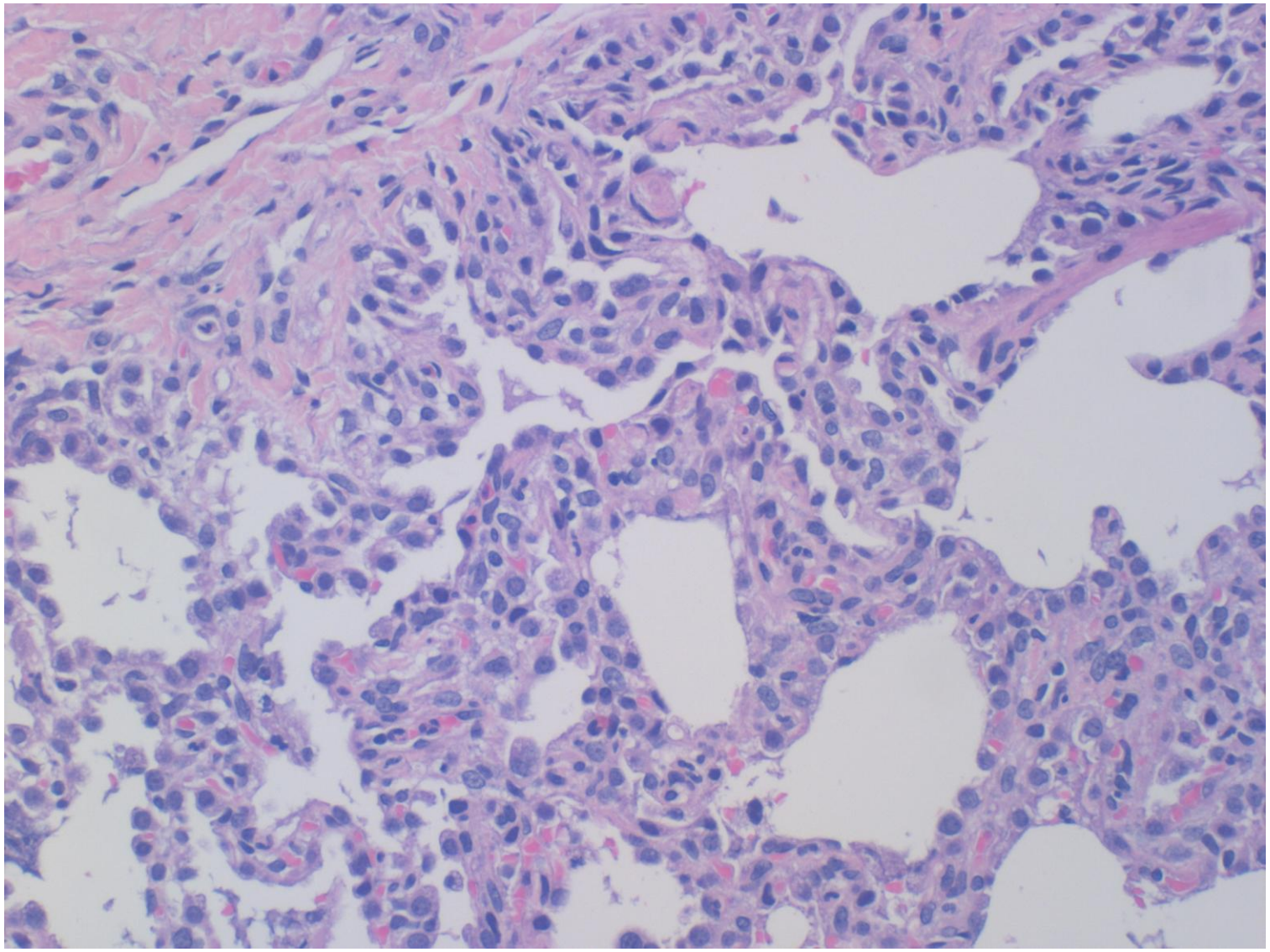
Interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activation of type II pneumocytes is seen along the mildly thickened alveolar septa. Exudation of alveolar macrophages is associated. H&E-2

Case 2



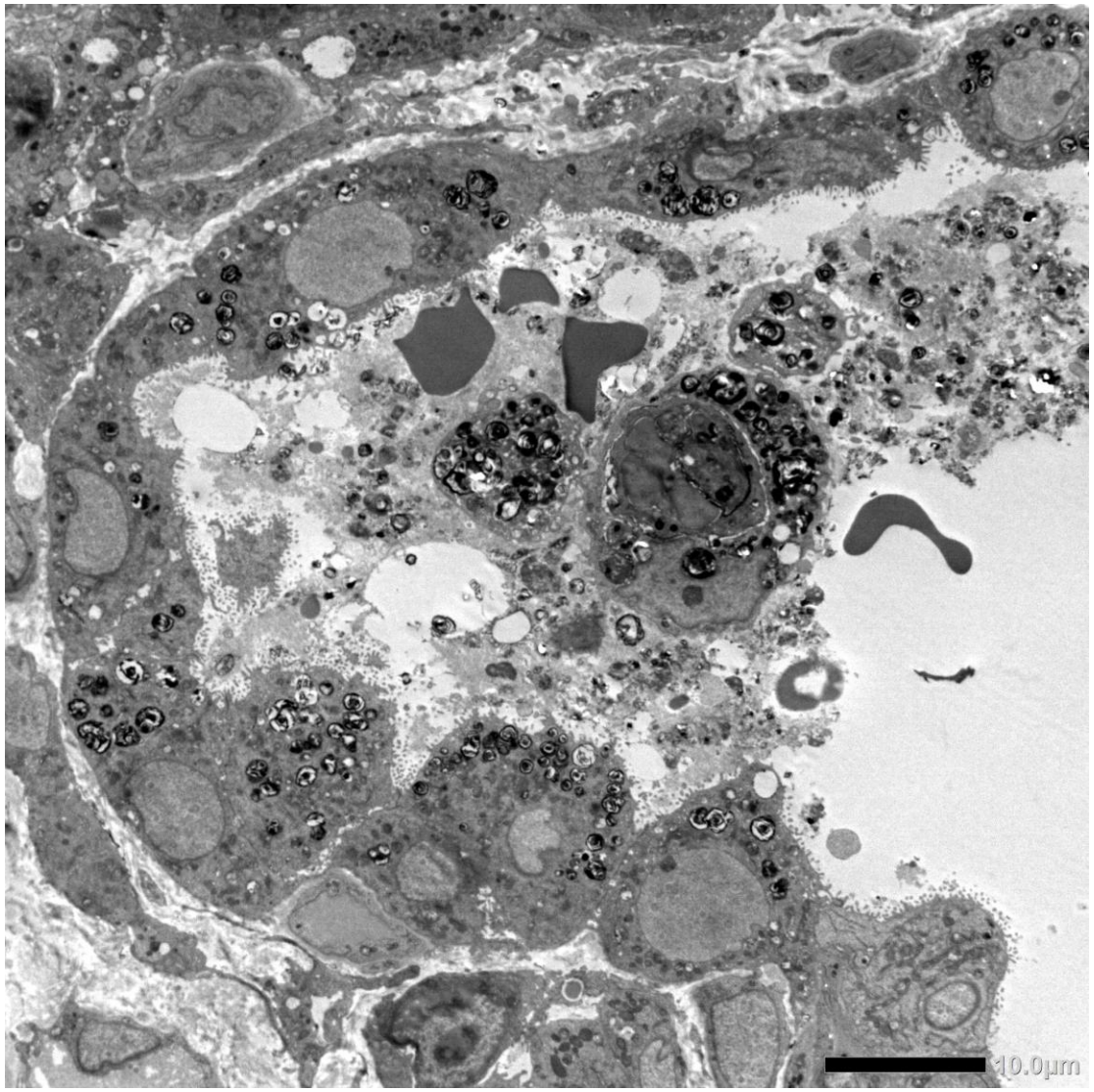
Interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activation of type II pneumocytes is seen along the mildly thickened alveolar septa, and focally occupy the collapsed alveolar lumen. Exudation of alveolar macrophages is associated. H&E-1

Case 2



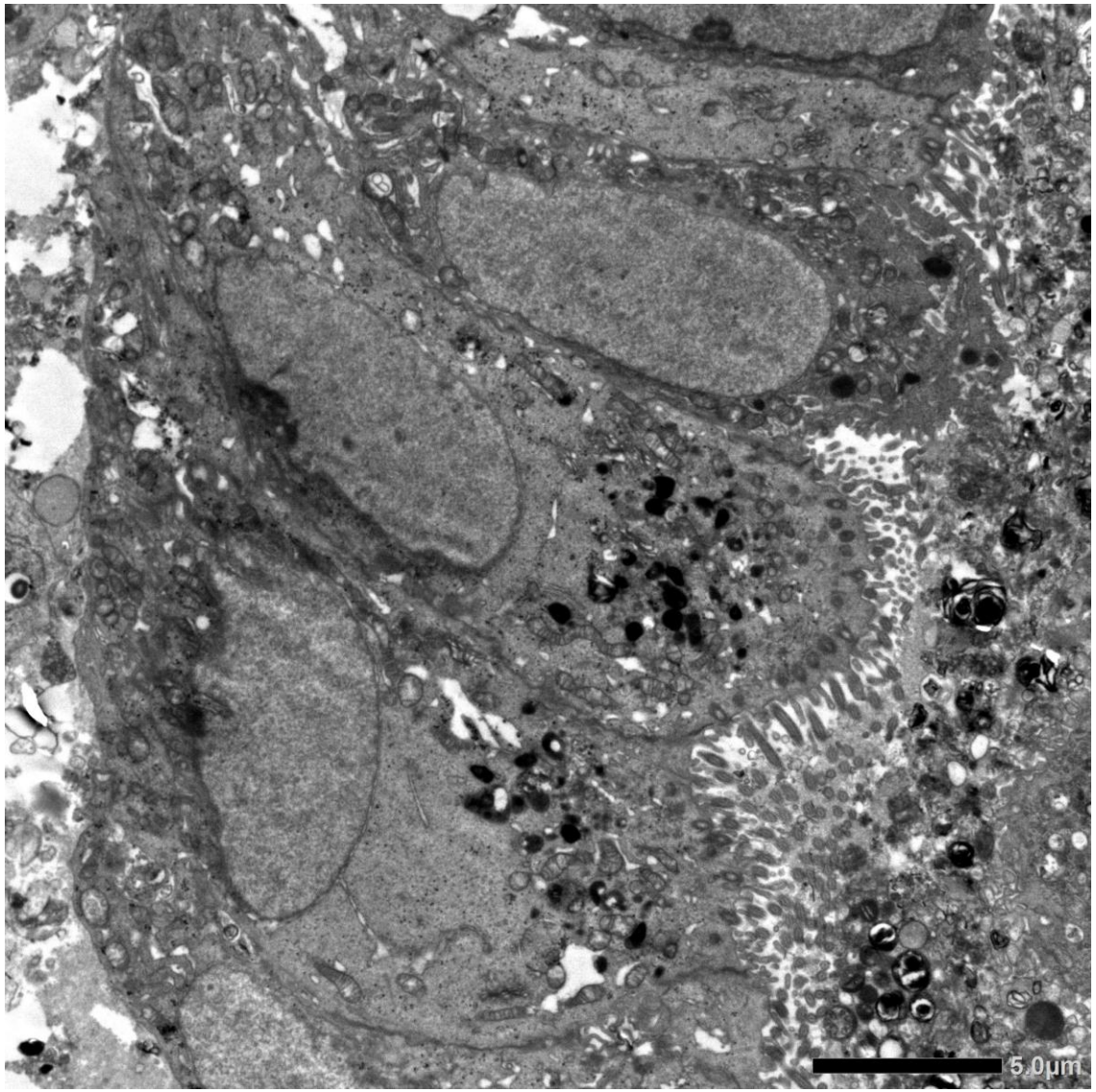
Interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activation of type II pneumocytes is seen along the mildly thickened alveolar septa. Capillary vessels increase in the fibrotic stroma. H&E-4

Case 2



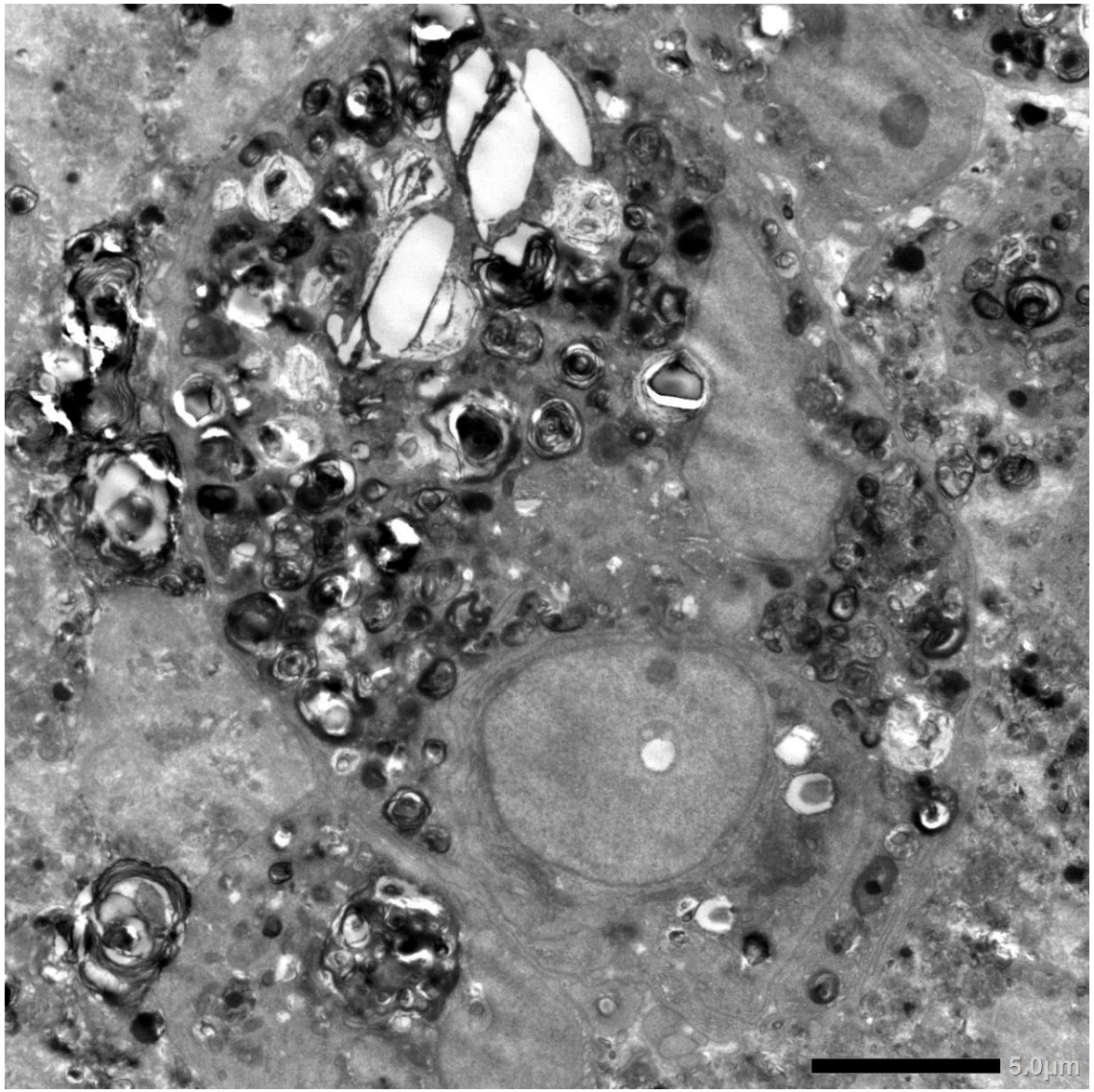
Ultrastructure of interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activated type II pneumocytes line along the mildly thickened alveolar septa. Exfoliated type II pneumocytes are seen the alveolar lumen. EM-1

Case 2



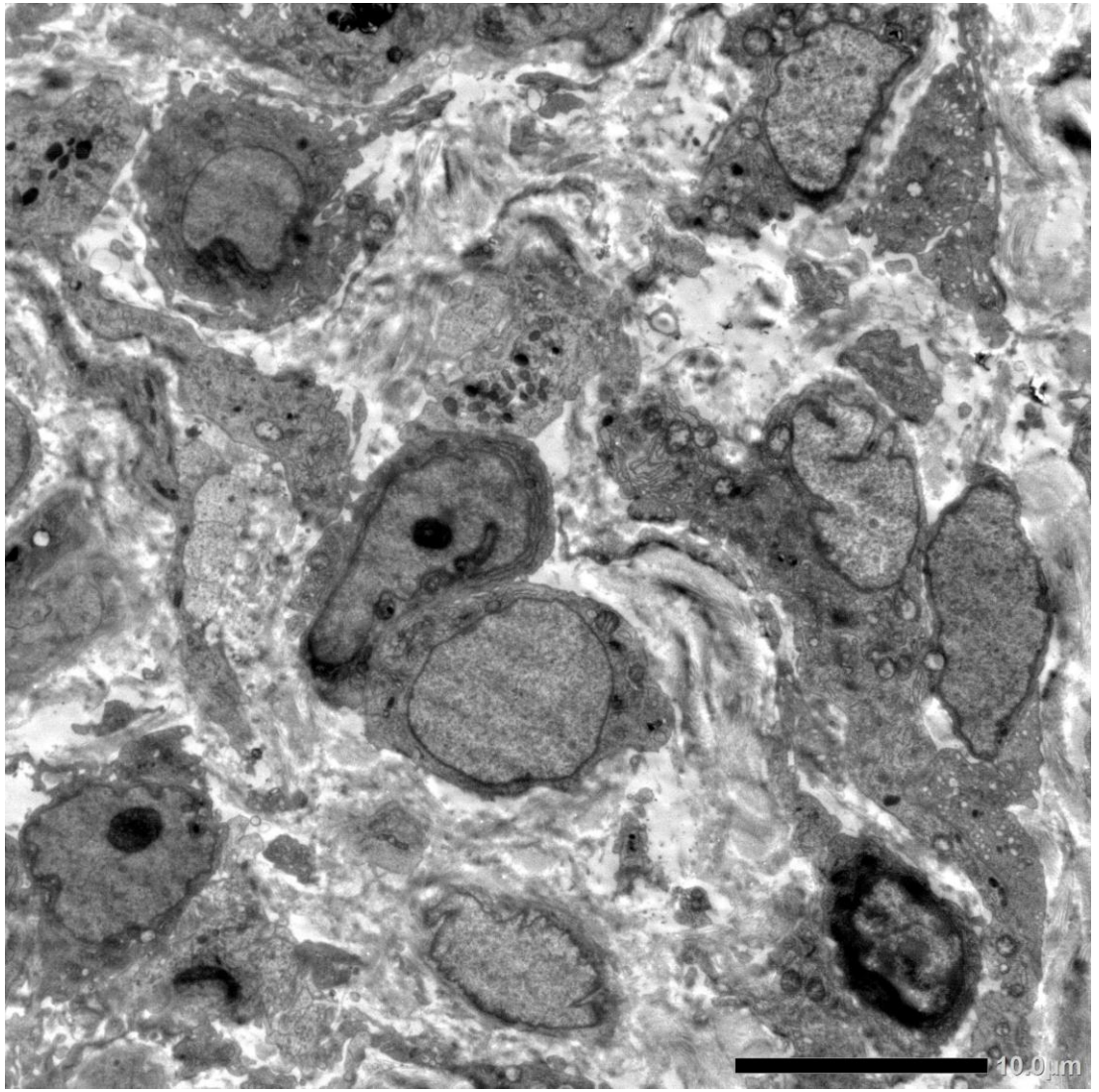
Ultrastructure of interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activated type II pneumocytes line along the mildly thickened alveolar septa. Microvillous surface is evident. EM-2

Case 2



Ultrastructure of interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, activated type II pneumocytes contain numerous lamellar bodies and deposits of cholesterin crystals. EM-3

Case 2

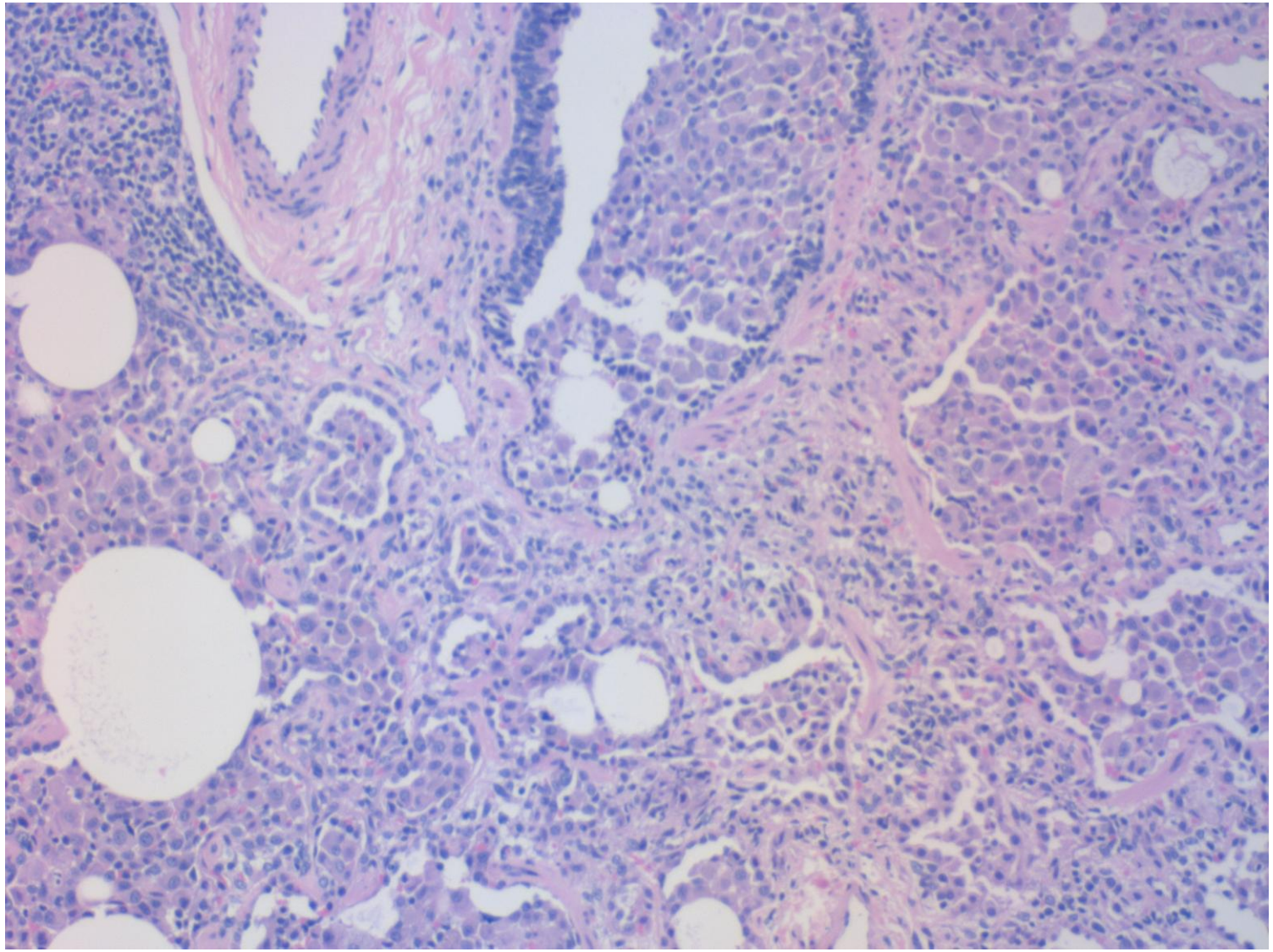


Ultrastructure of interstitial pneumonia associated with 21 trisomy (a 3-month-old girl). In this slowly progressive form of chILD, the alveolar lumen is lined by both type I and activated type II pneumocytes. In the mildly thickened edematous alveolar septum, fibroblastic cells are scattered. EM-4

Case 3: A 1-year-old girl

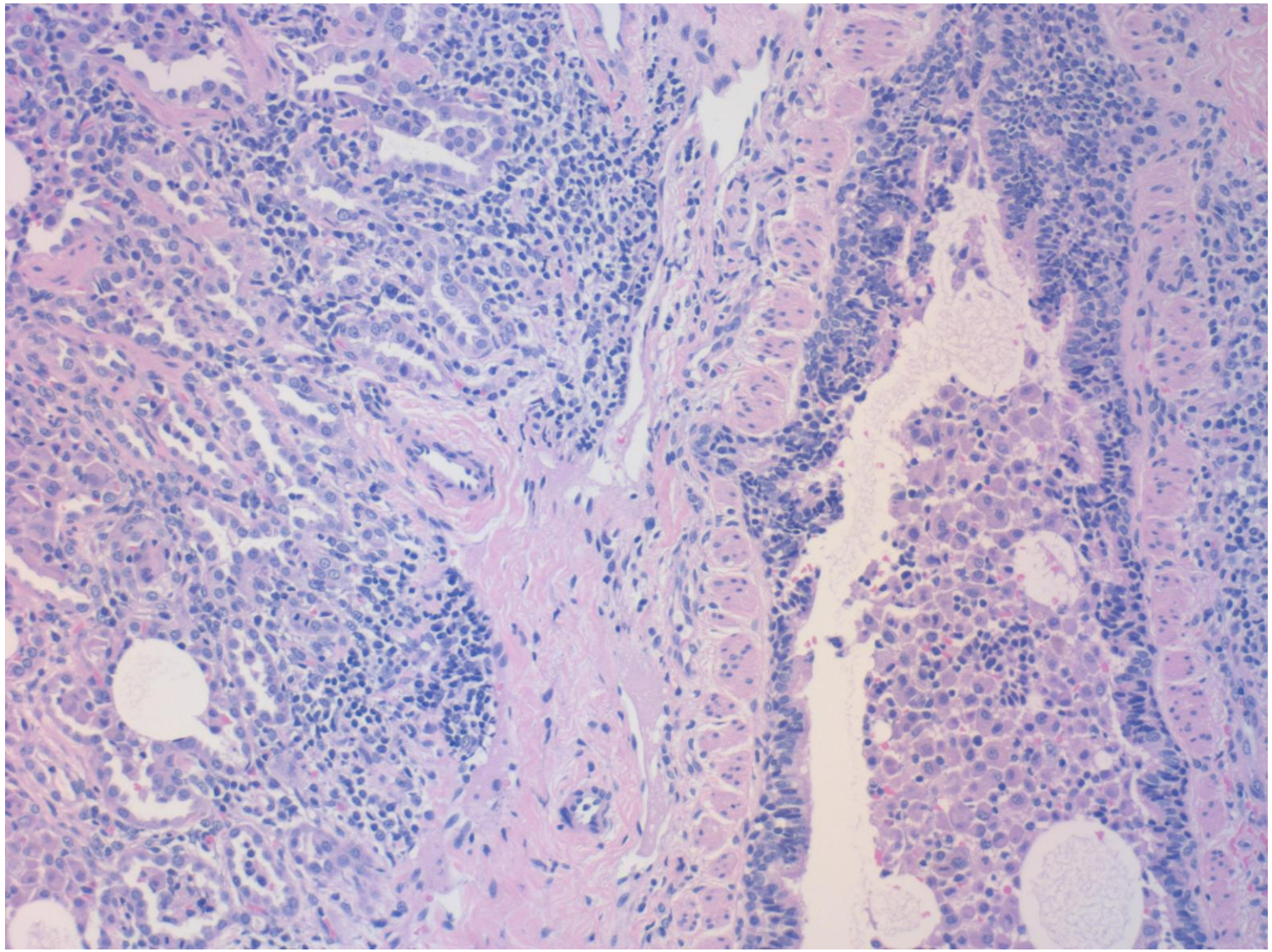
A newborn girl suffered from cytomegalovirus pneumonia 1 month after birth. Thereafter, elevation of KL-6 persisted and chest CT revealed bilateral ground-glass opacity. Interstitial pneumonia was clinically suspected, and VATS biopsy was taken 1 year after birth. The diagnosis should be desquamative interstitial pneumonia

Case 3



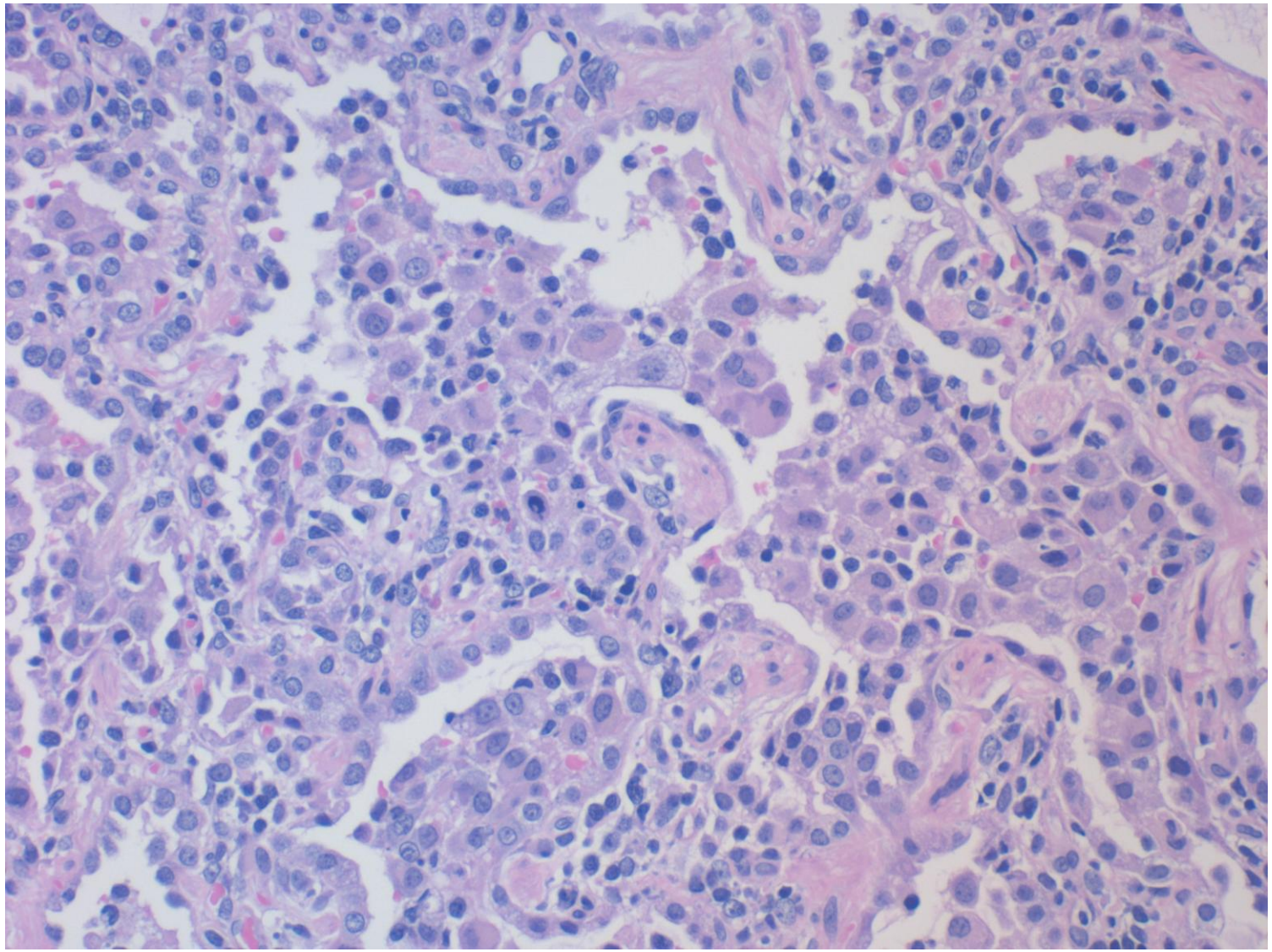
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activation of type II pneumocytes seen along the mildly thickened alveolar septa show marked desquamation into the alveolar lumina to reveal desquamative interstitial pneumonia. Desquamation is also seen in the bronchiolar lumen. H&E-1

Case 3



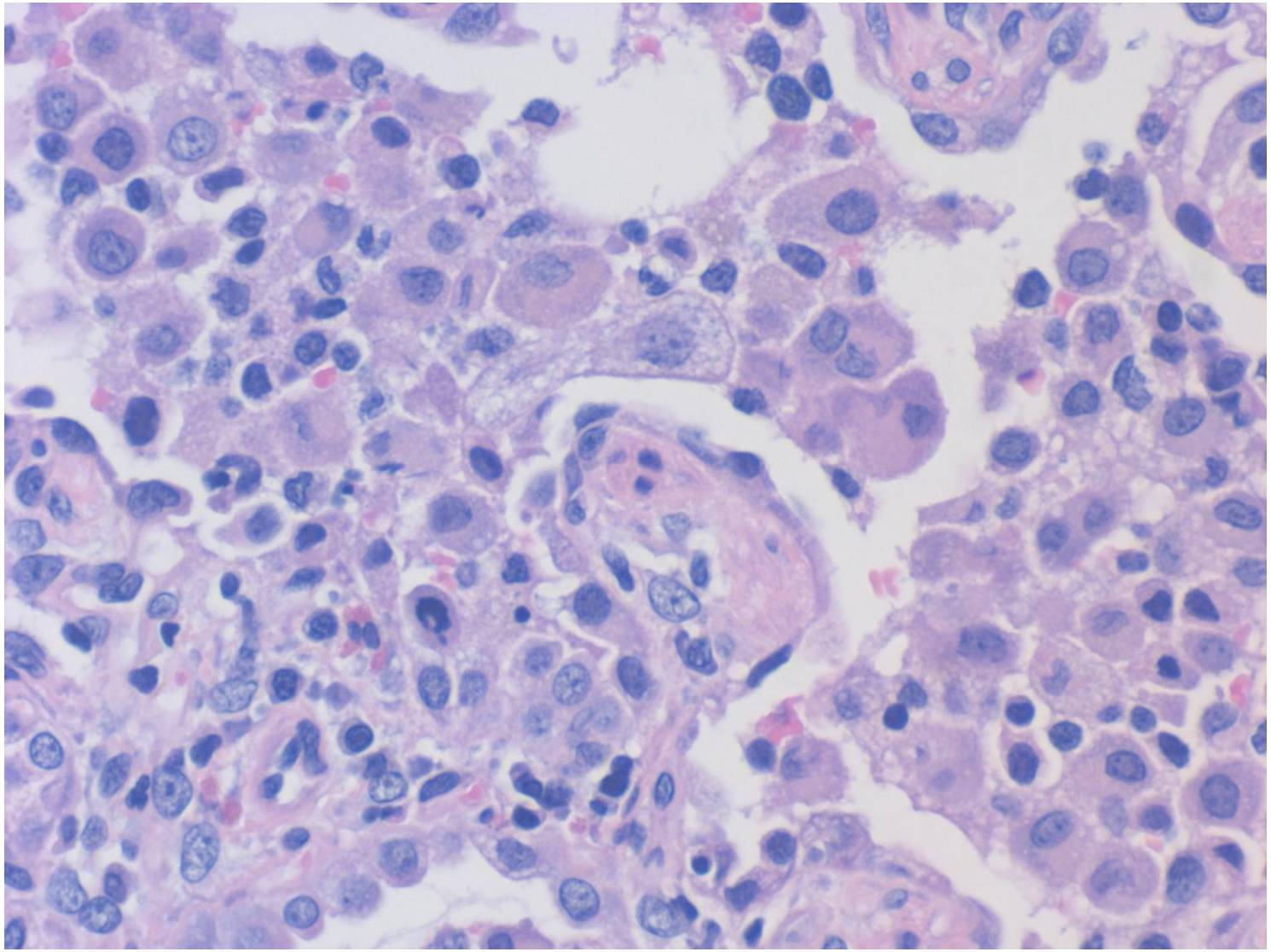
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activation of type II pneumocytes is seen along the mildly thickened alveolar septa. Mild stromal lymphocytic infiltration is associated. Desquamation is observed in the bronchiolar lumen. H&E-2

Case 3



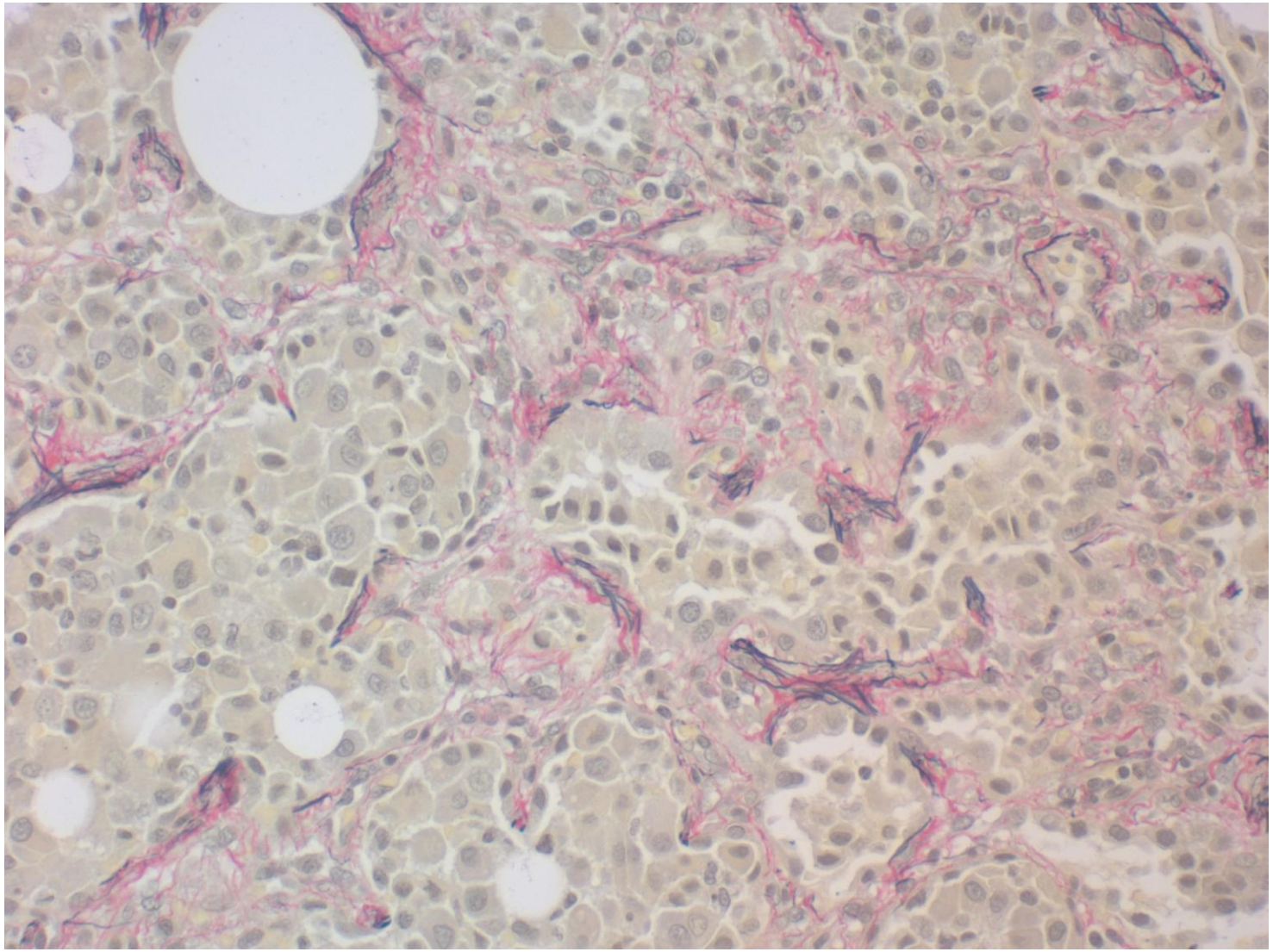
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activation of type II pneumocytes seen along the mildly thickened alveolar septa show marked desquamation into the alveolar lumina to reveal desquamative interstitial pneumonia. H&E-3

Case 3



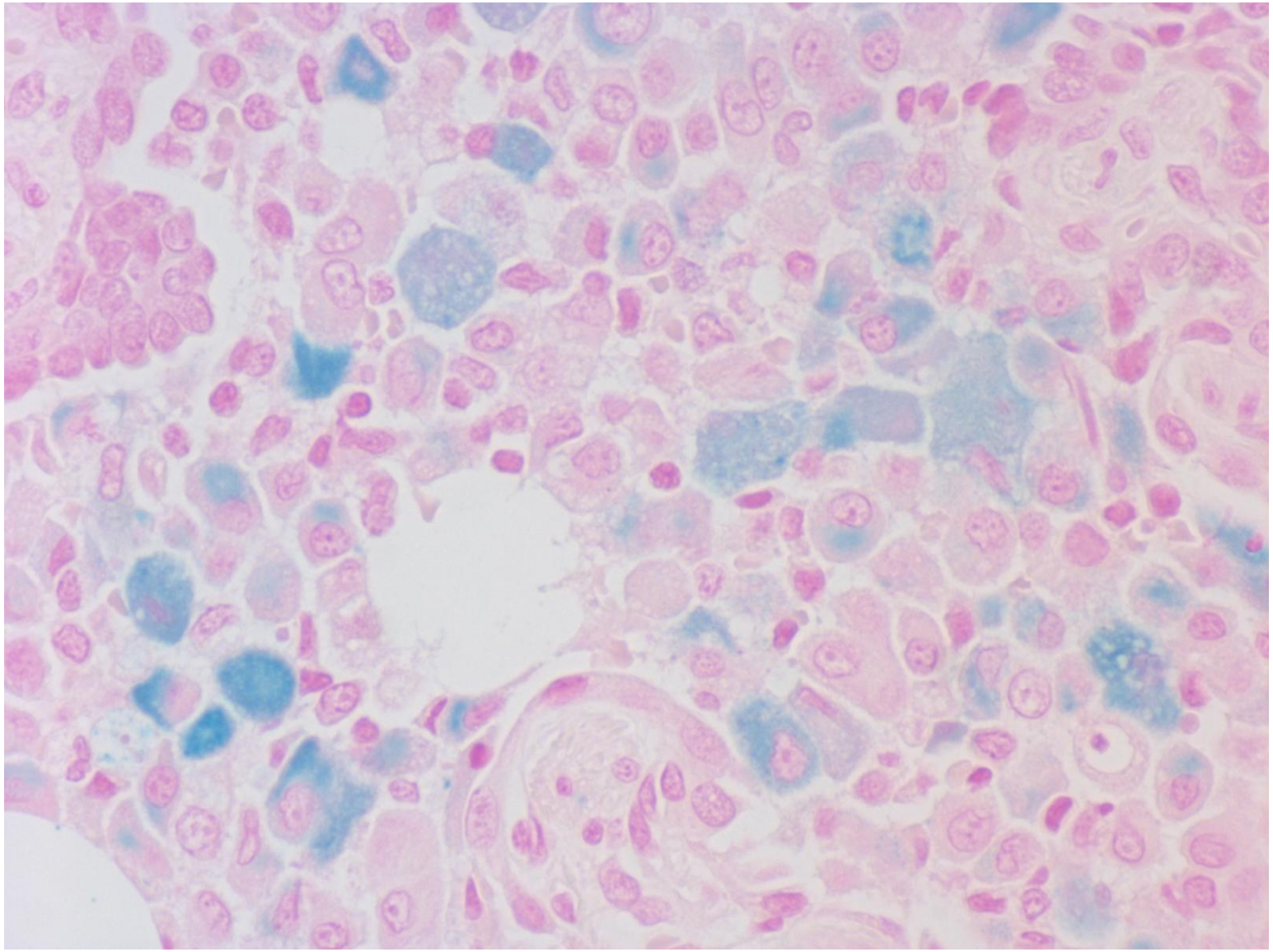
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activation of type II pneumocytes seen along the mildly thickened alveolar septa show marked desquamation into the alveolar lumina to reveal desquamative interstitial pneumonia. H&E-4

Case 3



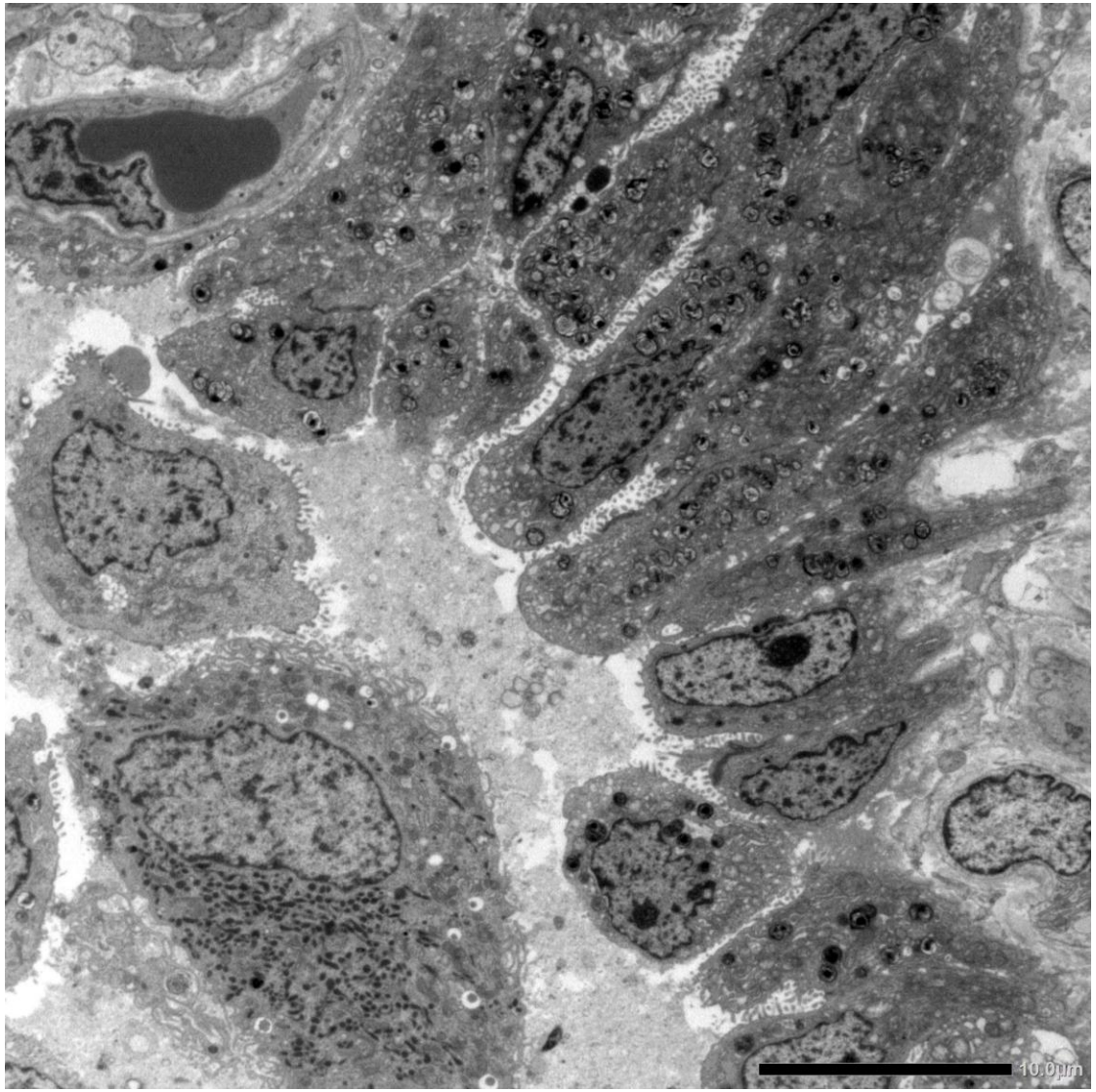
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activation of type II pneumocytes seen along the mildly thickened alveolar septa show marked desquamation into the alveolar lumina. The elastic fibers appear to be retained. EVG

Case 3



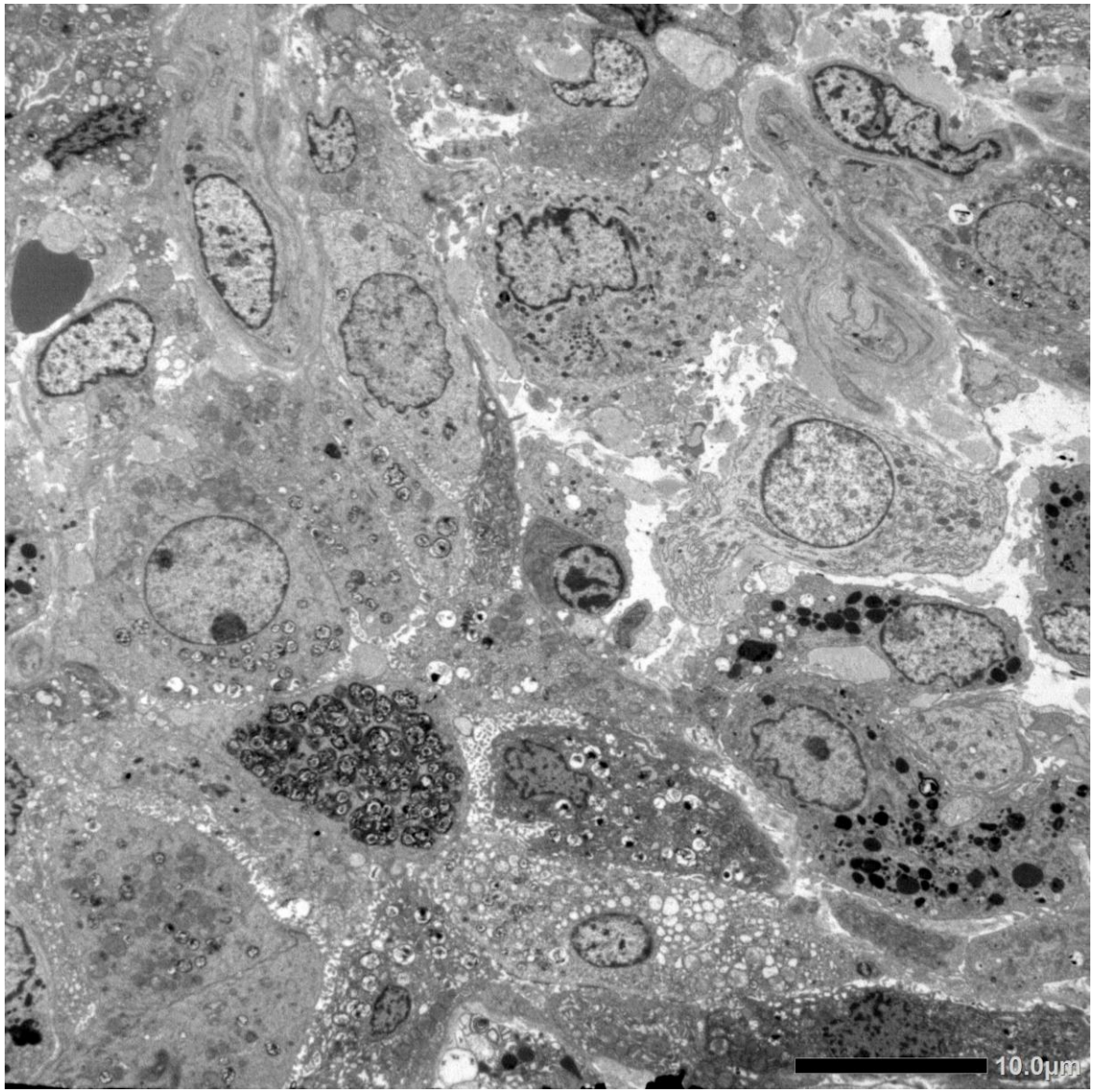
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activated type II pneumocytes are desquamated into the alveolar lumina, together with blue-stained hemosiderin-laden alveolar macrophages. Berlin blue

Case 3



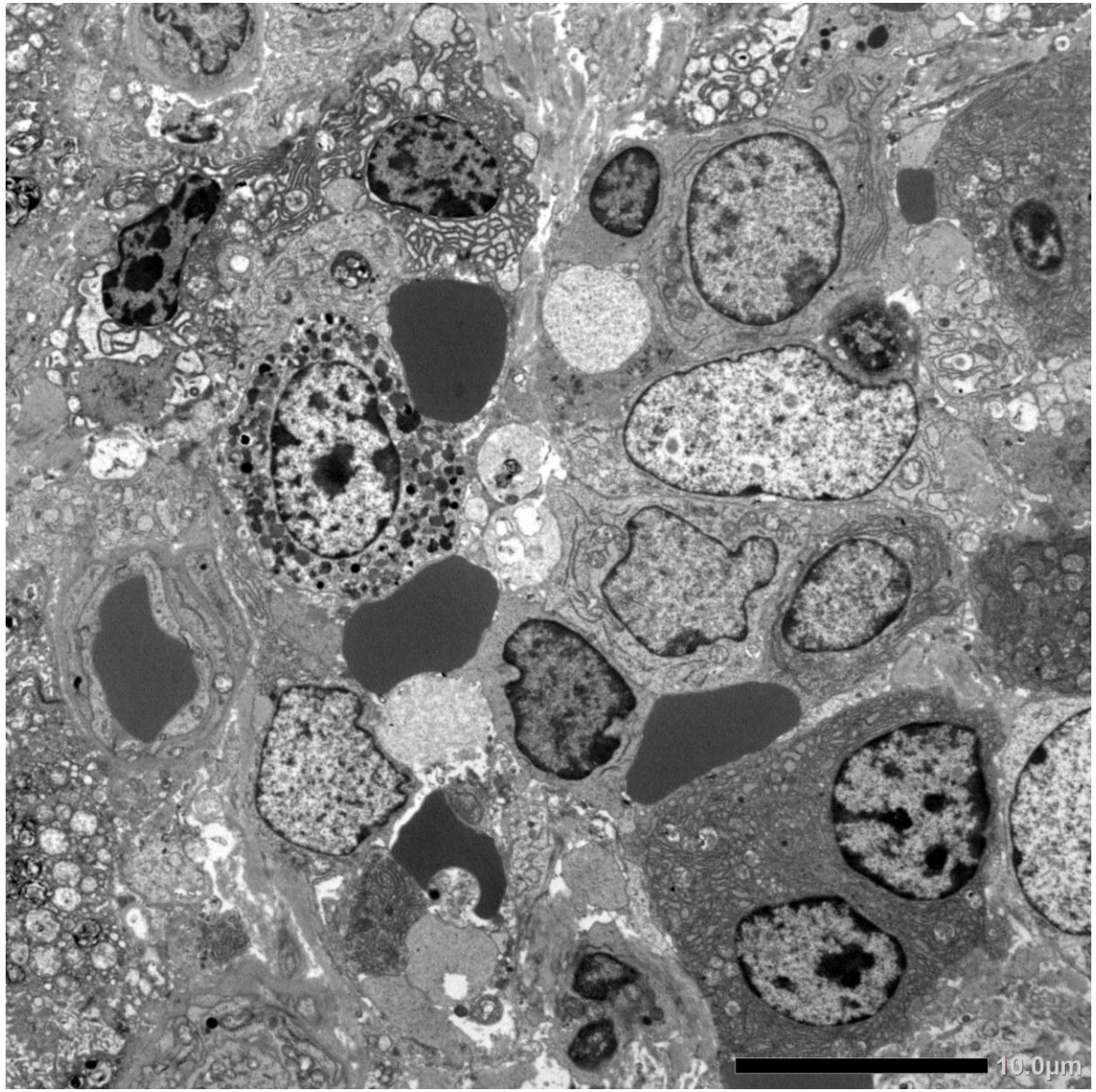
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, activation of type II pneumocytes is evident along the alveolar septum. Macrophages are desquamated into the alveolar lumen. EM-1

Case 3



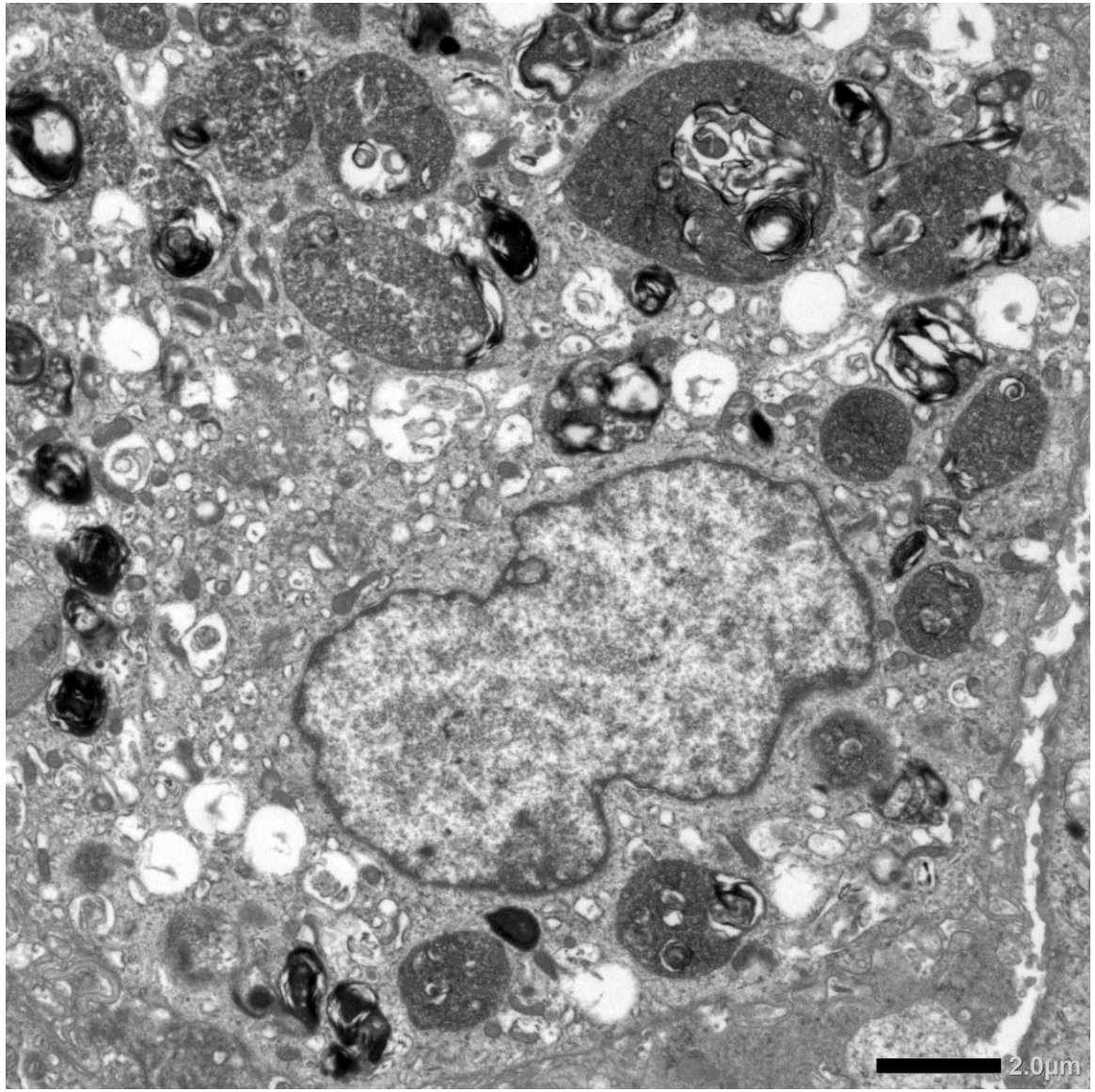
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, alveolar lumen is filled with desquamated type II pneumocytes with lamellar bodies and activated macrophages. EM-2

Case 3



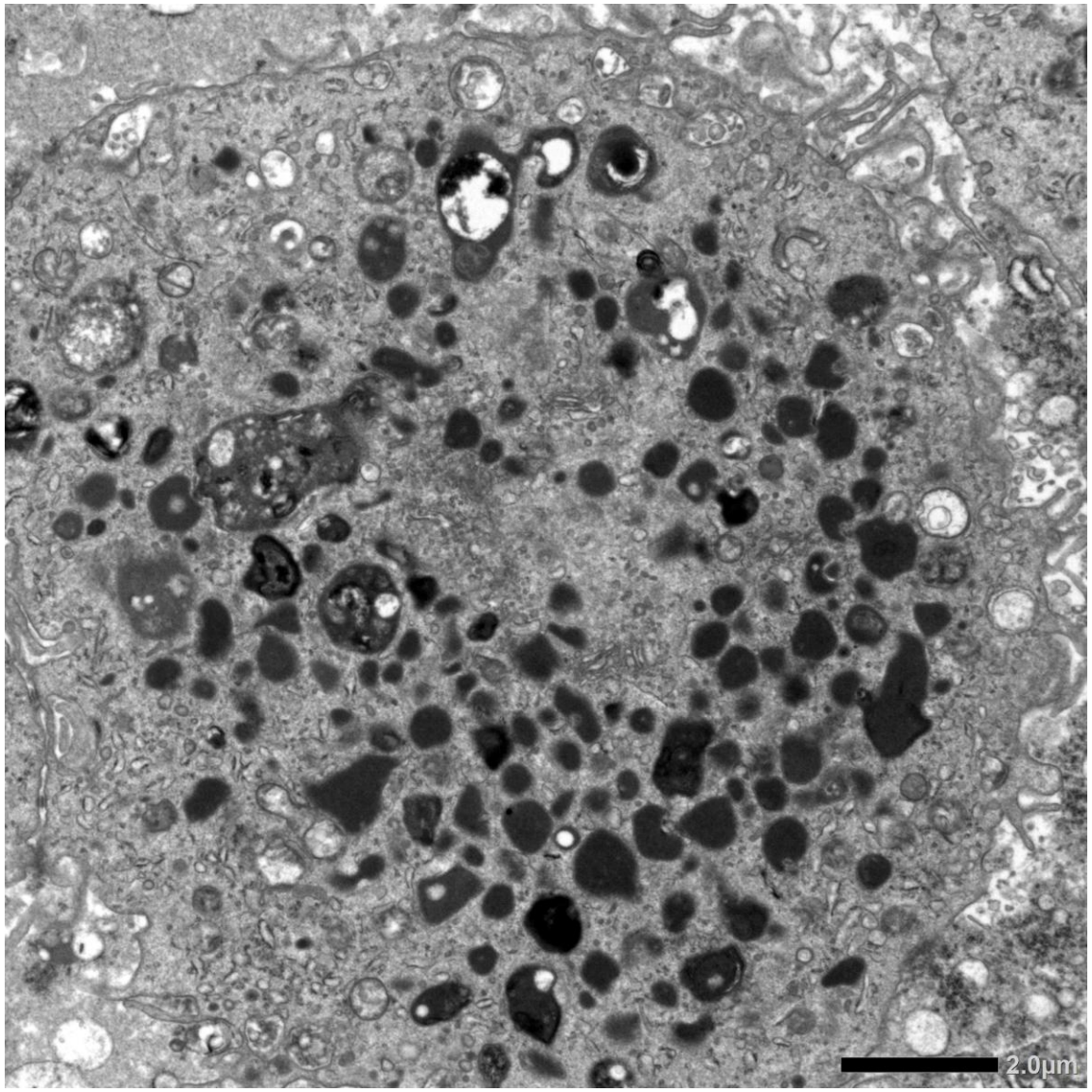
Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, alveolar lumen is filled with desquamated type II pneumocytes with lamellar bodies, red cells and activated macrophages. EM-3

Case 3



Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, a desquamated macrophage phagocytizes lamellar bodies in enlarged lysosomes. EM-4

Case 3



Interstitial pneumonia seen in a 1-year-old girl. In this slowly progressive form of chILD, a desquamated macrophage phagocytizes electron-dense hemosiderin granules in lysosomes. EM-5